Basic Pediatric Echocardiography

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BASIC PEDIATRIC ECHOCARDIOGRAPHY

2014

Mosul

Iraq

بسم الله الرحمن الرحيم

Dectated To

My Mother

Wife

Zeena,Zahra,Sura,Abdullah

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Early heart chambers

- Proliferation of the connective tissue precursors in the endocardium cause deposition of the future fibrous skeleton, which partitions the early heart.
- Proliferation of the myocardium lays down the muscular layer of the contractile chambers of the developed heart.
- The chambers of the early heart are as follows:

Sinus venosus (SV)	This is the *collecting compartment* of the heart. Oxygenated blood from the placenta and deoxygenated blood from embryonic tissues are mixed in the sinus.
Primitive atrium (PA)	This is the compartment destined for further partition to the definitive atria.
Primitive ventricle (PV)	This is the compartment destined for further partition to the definitive ventricles.
Bulbus cordis (BC)	This will contribute to the pulmonary trunk and aorta, along with the truncus arteriosus.
Truncus arteriosus (TA)	This will contribute to the aortic arches.
Fibrous skeleton (FS)	This is the area of connective tissue proliferation, site of future valves.

- A longitudinal section through the heart tube shows the arrangement of the early chambers(figure1.1).

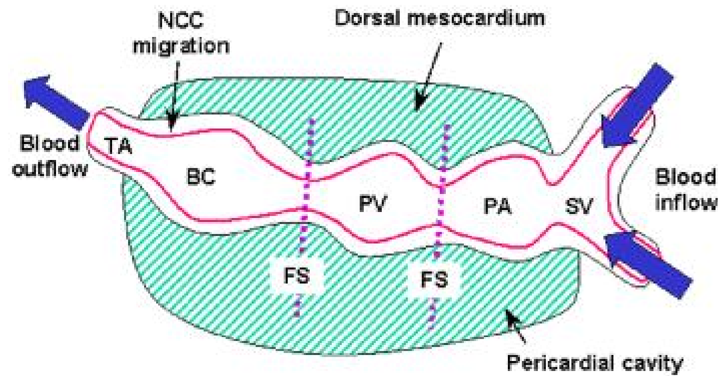


Figure 1.1

SV - sinus venosus; PA - primitive atrium; PV - primitive ventricle; BC - bulbus cordis;TA - truncus arteriosus; FS - fibrous skeleton; NCC - neural crest cell

- Neural crest cell migrate into the truncus arteriosus and bulbus cordis, where they contribute to the *aortico-pulmonary septum*.

The first dorsal fold

- Apoptosis in the dorsal mesocardium will allow movement of the early heart tube within the pericardial cavity(figure2.1).

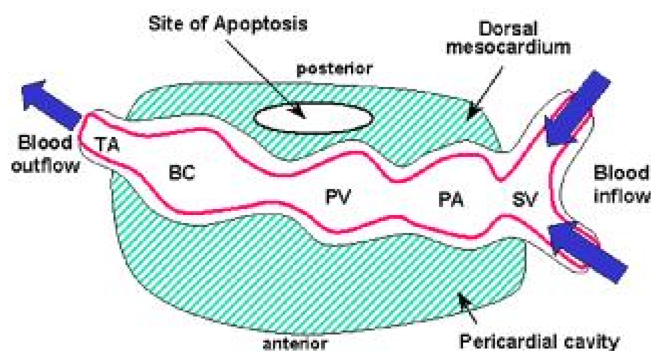


Figure 2.1

- The first dorsal fold brings the inflow and outflow trunks in an adjacent position at the superior aspect of the developing heart.
- Conceptually, the primitive ventricle can be seen as a fulcrum around which the heart tube folds.(figure3.1)

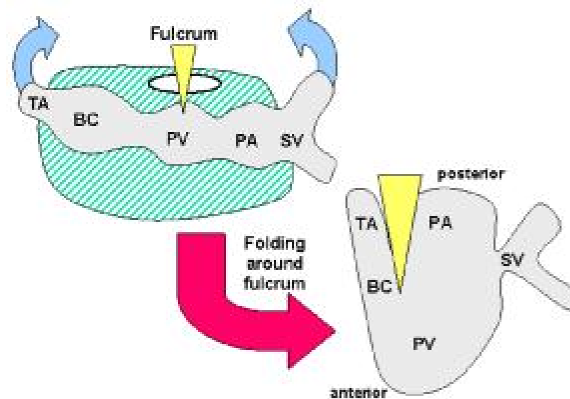


Figure 3.1

§ The dorsal fold forms the two pericardial sinuses and places the *fibrous skeleton* in a single plane(figure4.1).

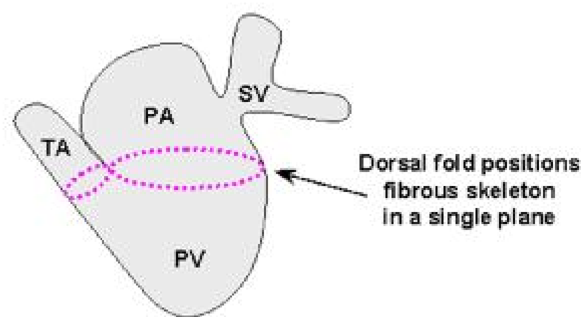


Figure 4.1

§ Inflow and outflow trunks are positioned posteriorly as a result of the dorsal fold(figure5.1).

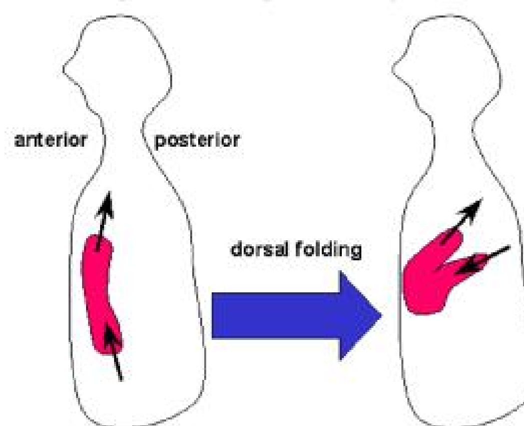


Figure 5.1

Endocardial cushion fusion

§ Endocardial cushions are areas of mesenchymal proliferation.
§ As described earlier, these represent areas of the fibrous skeleton forming between the atrium and ventricle.
§ Endocardial cushions serve two important functions:

ü They form a partition in the heart tube between the atrium (PA) and ventricle (PV), known as the AV canal. The resulting two channels represent sites for the future tricuspid and bicuspid valves.
ü provide a "scaffold" to which the interatrial septae and the interventricular septum will grow towards and fuse with(figure6.1).

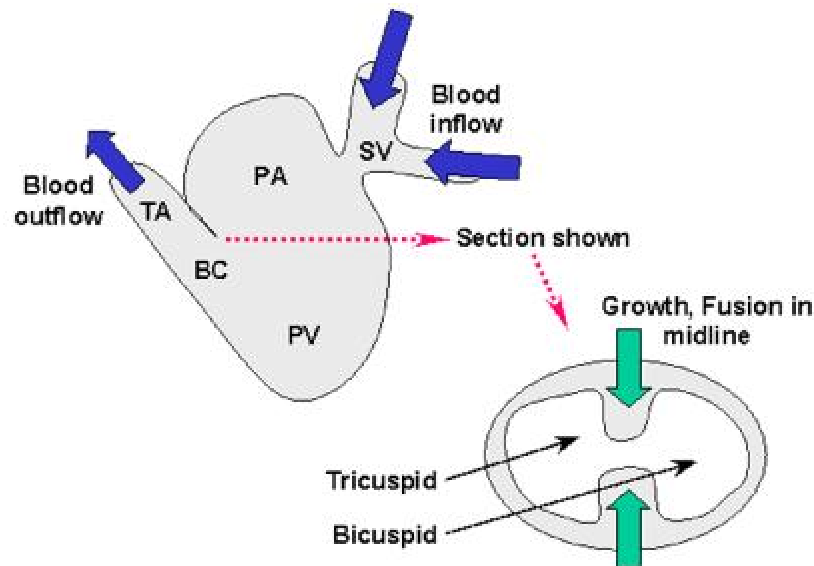


Figure 6.1

SV - sinus venosus; PA - primitive atrium; PV - primitive ventricle;
BC - bulbus cordis; TA - truncus arteriosus

§ Defects in endocardial cushion fusion are associated with trisomies 18 and 21 (Down's syndrome).
§ Since the scaffold for future growth is absent, ventricular septal defects are also common.
§ The resulting condition is referred to as *atrioventricular communis*. This anomaly is seen with varying degrees of severity - the most severe being characterized by 4-chamber communication.
§ The proliferation of the fibrous skeleton, combined with the fusion of the endocardial cushions will form the biscuspid (mitral) and tricuspid valves(figure7.1).

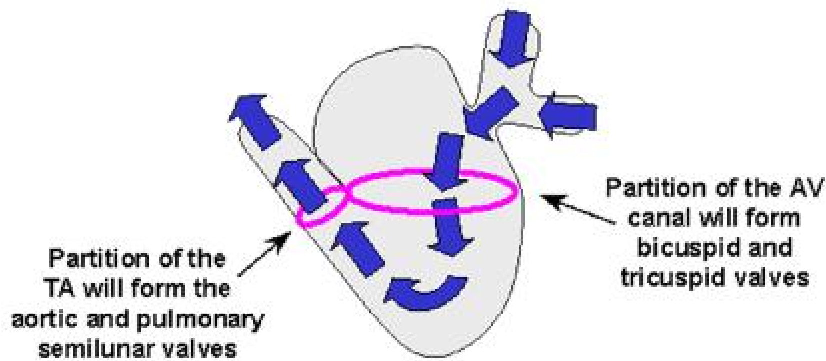


Figure 7.1

- *Valvular defects* can arise if endocardial cushion fusion does not partition the AV canal evenly.
- D- looping is a consequence of several changes:
  - *Dorsal folding* :The first dorsal fold forms an expanded primitive ventricle, referred to as the bulboventricular loop. This loop is subject to further changes, mainly of a hemodynamic nature.
  - *Ventricular growth* :Differential growth of the proximal ventricular tissue causes a counter-clockwise rotation of the folded heart tube. The site of ventricular growth marks the future left ventricle. Abnormal growth of the distal primitive ventricle causes clockwise rotation, an anomaly known as *dextrocardia*.
  - *AV canal partitioning* :The Atrio-Ventricular (AV) canal between the primitive atrium and ventricle has now been partitioned by the *fusing endocardial cushions*. The division serves to direct the blood preferentially through one channel.
  - *Shunting of venous return* :The development of the venous system causes an increase in right-sided venous return to the primitive atrium. Combined with the partitioning of the AV canal, the change in blood flow volume and directions assists in the outgrowth of the left ventricle.
- *Bulboventricular looping* is essential in the establishment of normal hemodynamic patterns,and thus normal development of septae and internal structures.

Interatrial septum formation

- The formation of the atrial septum must preserve the fetal shunting of oxygenated blood from the IVC across the atria to the systemic circulation, while allowing conversion to the adult blood flow pattern at birth.
- The stages of septal formation are outlined below.
- Legend for following figures:
  Blue arrows - direction of growth; Red arrow - direction of blood flow;
  ECC - endocardial cushion; RA - right atrium; LA - left atrium(figure8.1).
- *Septum primum* grows downwards from atrial wall. The opening between the growth edge of the septum and the fused endocardial cushions is known as *foramen primum*.
- Blood from the inferior vena cava will flow across the atria, through foramen primum to the systemic circulation.

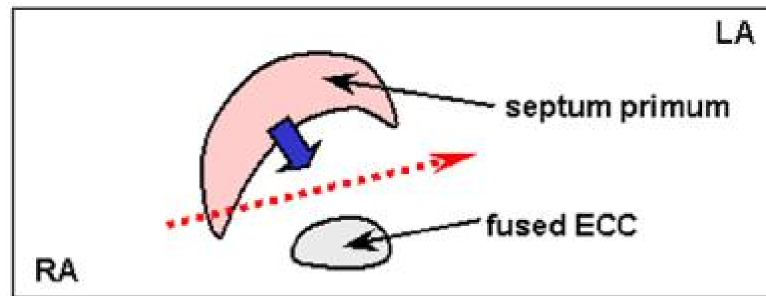
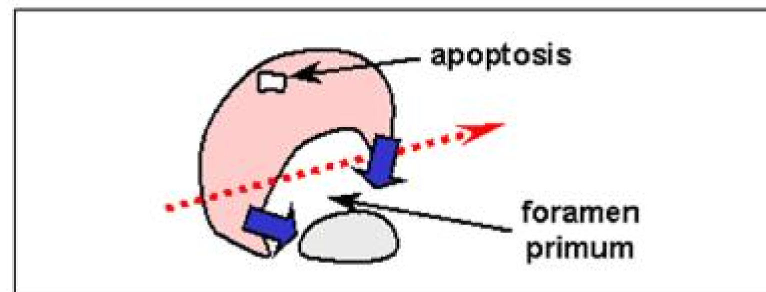
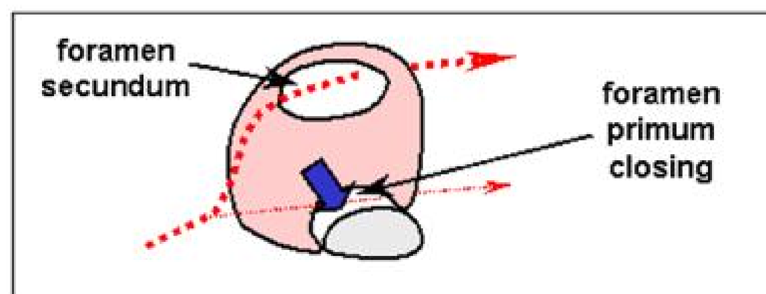


Figure 8.1

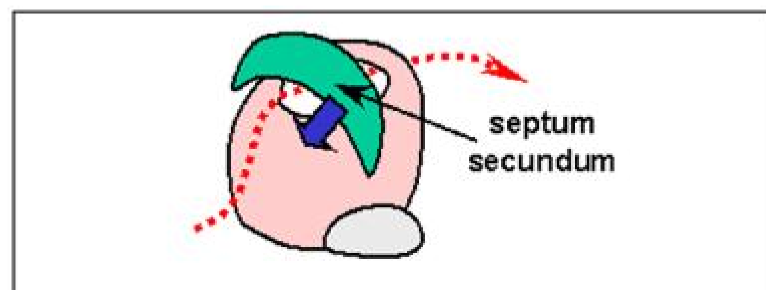
- As growth progresses, foramen primum is reduced in diameter. Apoptosis in the upper portion of the septum begins.



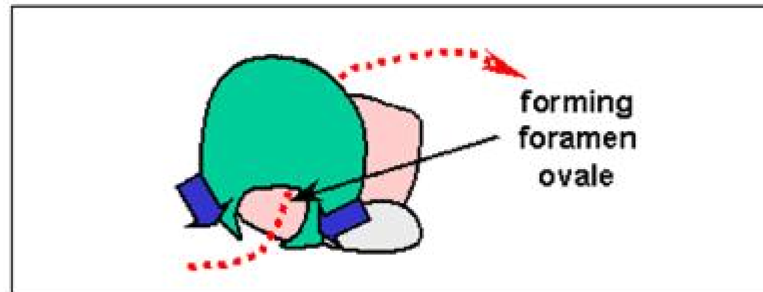
- The growth edge of septum primum now fuses to the endocardial cushion mass.
- Apoptosis in the superior aspect of *septum primum* forms a new opening, the *foramen secundum*.
- Any defect in the fusion of septum primum to the endocardial cushion is referred to as a patent foramen primum. These are usually slight and of no physiological importance.



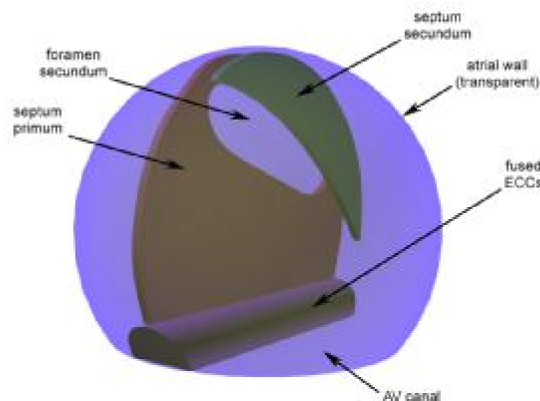
- The *left valve of the sinus venosus* begins growth downwards as *septum secundum*.



- Growth and fusion of septum secundum to the endocardial cushions leaves an opening, the *foramen ovale*.
- The superior-most aspect of septum primum apoptoses, freeing this edge from the atrial wall. Septum primum now acts as a valve for the opening in septum secundum.



- Seen in three dimensions (at approximately stage 4 above), only the atria have been shown in this perspective view for clarity.

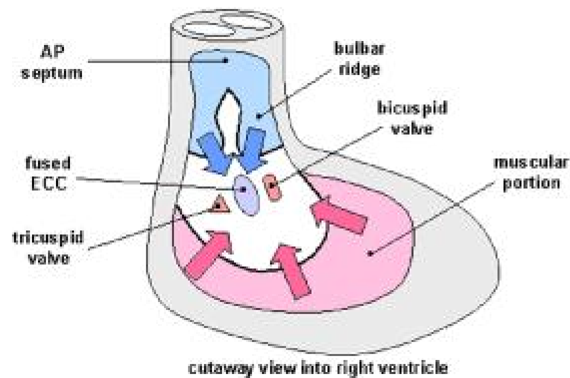


- After birth, the pressures generated in the left atrium press septum primum against foramen ovale, sealing the collateral blood flow off and establishing the separate pulmonary and systemic adult circulations.
- The *limbus of the fossa ovalis* represents the edge of the septum secundum fused with the septum primum.
- *Endocardial cushion fusion* is the critical first step to proper interatrial septum formation: the central mass of fused endocardial cushions is the "scaffold" to which the growing septum primum and secundum will fuse. Absence or irregularities of growth, or the failure to close of either septum primum or secundum will also produce *septal defects*.

Interventricular septum formation

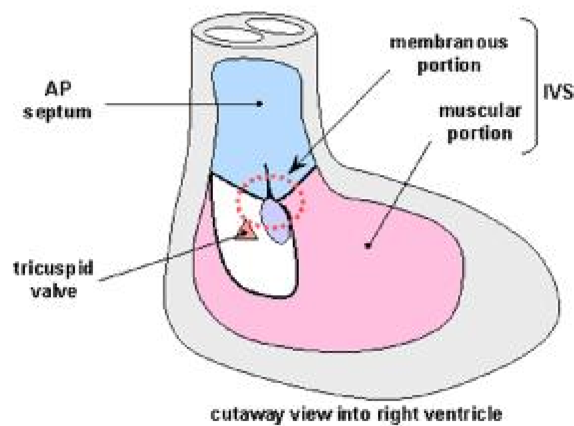
- The interventricular septum (IVS) serves as the *final event* in separating aortic and pulmonary *outflow* from the heart.
- The division between IVS formation and aortico-pulmonary (AP) septum formation is done in order to highlight important features of each event. *In reality, however, the two occur simultaneously*.
- The IVS is characterized by two parts, contributed by three separate structures:

- ü *a muscular portion* :The muscular wall grows upwards towards the fused endocardial cushions, separating the bicuspid and tricuspid valves (and thereby, in- and outflow).
- ü a membranous portion :The membranous portion of the IVS is contributed by the fused endocardial cushions, as well as the descending *bulbar ridges*.



AP - aorticopulmonary; ECC - endocardial cushion;
Blue arrow - direction of bulbar ridge growth; Red arrow - direction of ventricular growth

- § Once the interventricular septum is formed, the tricuspid and bicuspid valves are separated, thereby diving cardiac outflow into pulmonary and aortic streams. The membranous portion is outlined with a dotted red line.



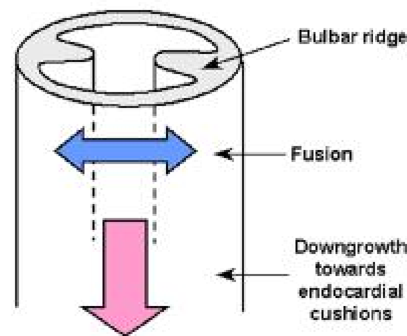
- § 25% of all ventricular septal defects occur in the membranous portion, *due to the complexity of fusing three separate components together*.
- § To summarize the origin of the interventricular septum:

IVS Part	Origin
muscular	• ventricular wall
membranous	• fused bulbar ridges • fused endocardial

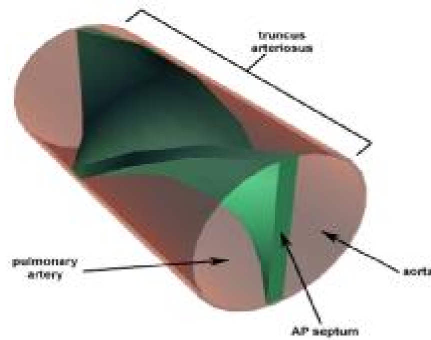
	cushions

Aortico-pulmonary septum formation

- The aortico-pulmonary (AP) septum arises within the *truncus arteriosus*. The septum results from the downwards growth and fusion of bulbar ridges, induced by invasion of neural crest cells.
- The AP septum serves to divide the ventricular outflow between the pulmonary artery and the ascending aorta.



- In addition to dividing cardiac outflow, the AP septum *contributes to the formation of the semilunar valves*.
- Anomalous formation of the AP septum can lead to several anomalies. *Eisenmenger's syndrome* occurs as a result of equal division of the truncus with an incomplete fusion of bulbar ridges distally.
- When the division of the AP septum occurs asymmetrically, the *tetralogy of Fallot* results.
- Once fusion has occurred, the AP septum has the following appearance (the tube has been made transparent to show the fused bulbar ridges).
- Note the symmetric division of the truncus arteriosus, and the 180-degree twist given to the septum.



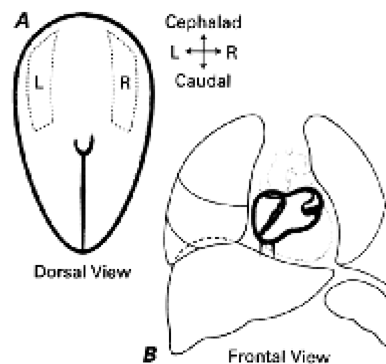
2-Cardiac development

The First Choice: Sidedness.
Morphogenesis of the Ventricles.
Formation of the Left Ventricular Outflow Tract and the Right Ventricular Inflow Tract.
Atrial Septation.
AV Canal and Ventricular Septation
Truncoconal Septation: The Formation of the Aortic and Pulmonary Trunks.

- In this review, embryologic phenomena-' are described and abnormal embryologic events that could contribute to the morphogenesis of congenital heart defects are suggested.

The First Choice: Sidedness

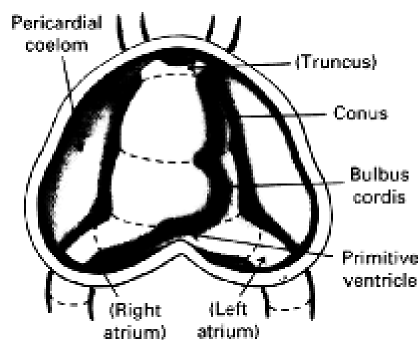
- The early vertebrate embryo at the primitive streak stage (about 15 days after fertilization in humans) is morphologically symmetrical, as shown in histologic studies .



L=left cardiogenic area……R=right embriogenic area…(R+L=cardiac primordial)

Situs solitus. A) Schematic dorsal view of the early chicken embryo at the primitive streak stage (17 hours of incubation). This stage corresponds to the end of the 2nd week after fertilization in the human embryo. The embryonic disc is symmetrical, with minimal organization. The cardiac primordia are located in front of the cephalic end of the embryonic disc. Later on, the ventral formation of the foregut will cause the cardiogenic areas to be located ventrally, on each side of the foregut (pharynx). B) The normal situs solitus viscera are histologically symmetrical at this early stage, but they have committed potentials to develop the asymmetric anatomic patterns shown in the lungs, atria (heart), liver,spleen, and intestines.

- The primordia of the cardiovascular system originate as clusters of paired, symmetrical mesenchymal cells in the coelomic mesoderm. Initially located on the *cephalad and dorsal aspect* of the embryo, they soon appear to migrate around the buccopharyngeal membrane of the forming foregut, and join at the midline of the ventral aspect of the embryo.
- The fused section of the cardiovascular primordia lies within the cephalad section of the originally undivided coelomic cavity, which soon defines a midline thoracic cavity (the pericardium).
- Structures within the pericardial cavity are defined embryologically as the heart.At first, the fused strands of cells of the primordia are arranged as a straight structure called the *straight tube heart, which traditionally has been described in terms of prospective segments: atria, primitive ventricle, bulbus cordis, conus, and truncus* .



The ventral view of the 4- to 8-somite embryo shows the formation of the straight tube heart within the pericardial coelom. The caudal (atrial) and cephalic ends of the primitive heart maintain the separation in 2 nearly symmetrical structures: the right and left atria; and the right and left 1st aortic arch. Embryologists have been able to identify the prospective regions of the heart at this early stage, as shown.Only the atria maintain strict derivation from just ONE cardiogenic area. The other segments combine derivatives from both the right and the left cardiogenic primordia.

- The heart at this stage has no inner circulation of blood, but soon becomes hollow (tubelike). *The mesenchymal cells forming the straight tube can be visualized as continuously migrating from the dorsal and cephalic aspects of the embryo toward the intrapericardial structure.*
- At the caudal extreme, the intrapericardiac structure meets the venous system, which, at this stage, comprises mainly the omphalomesenteric veins. From this caudal extreme, too, cells actively reproduce and migrate into the cardiac tube.

Developmental Implications.

- The 2 cardiogenic areas are provided with intrinsic differential growth characteristics and are the determinants of sidedness, which is the first and fundamental choice that the primitive heart makes.
- The cardiac structures originate by fusion of the 2 primordia, with the exception of the caudal extreme: *the right atrium receives only right-sided cardiogenic cells, and the left atrium, left-sided cardiogenic cells.*

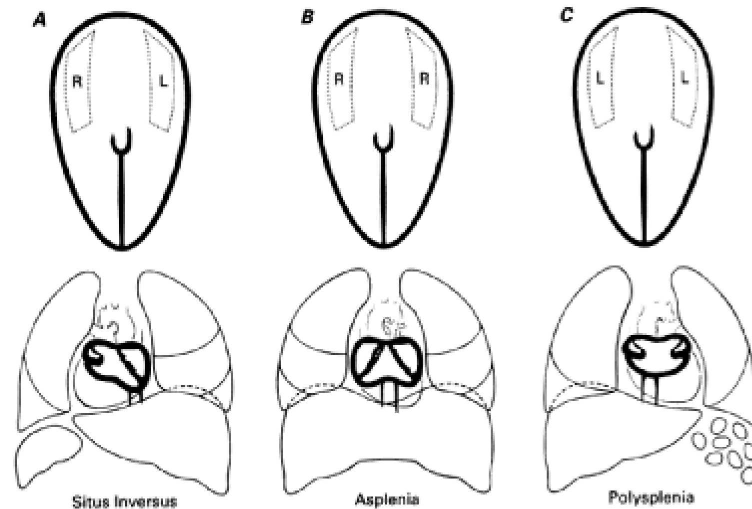
- The normal heart "decides" its visceral situs (pattern of asymmetry) at a very early stage of development . Fundamental anomalies, such as situs inversus and situs ambiguus of the dexter (asplenia) and sinister (polysplenia) types, might be explained by the developmental errors .

Morphogenesis of the Ventricles

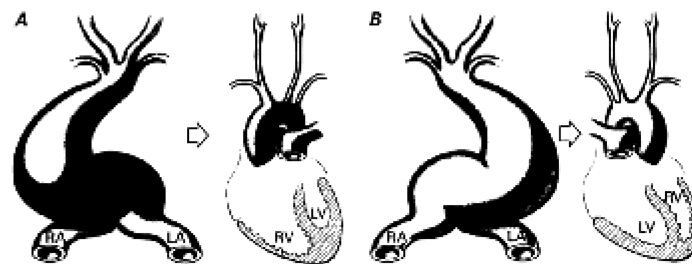
- Cardiac Looping :The embryonic heart acquires morphologic asymmetry, under the impulse of continuous cellular migration and multiplication, by looping the bulboventricular segments inside the pericardial cavity.
- Differential migration and multiplication of cardiac primordial cells could be the original reasons for normal evolution of the straight heart into a *dextroverted (or rightward) loop*, *called D-loop*.
- Experimental evidence has shown that alteration of the normal differential growth pattern may result in a leftward loop, or Lloop.The direction of looping appears to establish irreversibly the relationship between the ventricles (whose primordia, originally situated in series, become positioned side-by-side) and the already determined situs of the atrial cavities."' The looping of the bulboventricular tube leads the bulbus cordis (prospectively, the right ventricle) to the right (D-loop) or to the left (L-loop) of the initially caudal segment, which is the primitive ventricle (prospectively, the left ventricle).
- Additionally, the pattern of looping determines the relationship between the 4th and the 6th aortic arches. Because the 4th aortic arches are more anterior than the 6th, the looping of the bulboventricular segments will also establish a slight, but definite and irreversible shift to the right (D-loop) or the left (Lloop)of the anterior aortic arches (4th) with respect to the posterior structures. Such consequential morphogenetic definition results in a permanent relationship between the distal aortic and pulmonary trunks that will be affected neither by the eventual inner septation of the truncoconal structures nor by the shifting toward the midline of the caudal extreme of this tube (see section on truncoconal septation). The correlation between the position of the ventricles and the aortopulmonary relationship at their cephalad extreme is the basis for the so-called loop rule of concordance of the arterioventricular morphologies.

Developmental Implications

- Bulboventricular looping (to the right or the left) is a *morphogenetic process*, independent of the one that leads to the cardiac situs determination.
- Whereas the positions of the right and left atria are *determined by a certain cardiac (and visceral) situs*.
- The side-to-side relationship that develops between the ventricular cavities and the aortic and pulmonary trunks is decided by the *pattern of looping*.
- Four basic combinations are possible: *situs solitus with D looping*, *situs solitus with L- looping*, *situs inversus with D looping*, *or situs inversus with L- looping*.
- *D-looping* causes the caudal segment of the loop (the primitive ventricle)to lie to the left of the cephalad segment of the bulboventricular loop (bulbus cordis, prospectively the right ventricle). The embryologic conotruncus will also be displaced to the right, since it is serially connected with the bulbus cordis. (with rare exceptions the aorta is correspond to the same site of right ventricle)
- L-looping leads to a mirror-image arrangement of the bulboventricular components and their derivatives, in the frontal plane.

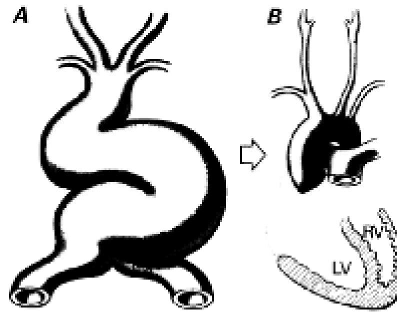


Classic experiments in chicken embryos have shown the many potentials of the right and left cardiogenic areas, including all types of variations in the positions of the atria, which are the only sections of the heart that maintain the individual nature of each cardiogenic area. A) Transplanting the right cardiogenic area in the left location and vice versa leads to the development of a mirror image, or situs inversus. B) Similarly, an embryo given 2 cardiogenic areas with right potentials will develop situs ambiguus dexter or asplenia syndrome (2 right atria). In naturally occurring congenital defects of the cardiogenic areas, the 2 entire hemisomas of the embryo commonly are affected, leading to accompanying anomalies of the situs of all viscera. C) When an embryo is transplanted with 2 cardiogenic areas having left-sided potentials, situs ambiguus sinister or polysplenia will develop.



The morphogenetic implications of bulboventricular looping are illustrated, showing the positions of the ventricles and of the cephalad extreme of the great vessels. The cephalic portion of the aorta shifts to the right in D-loop (A), and to the left in L-loop (B).LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle

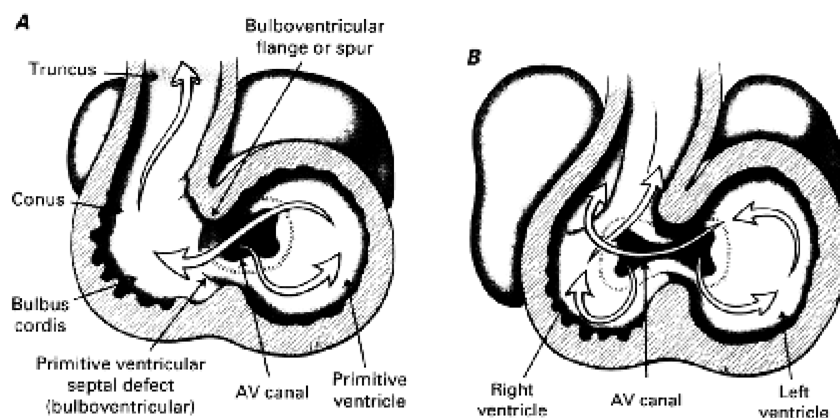
- During the last 2 decades of experimental embryology and clinical anatomy studies, it has become apparent that other types of grossly atypical looping are possible. These variants lead to malformations in which the ventricular anatomy does not conform to the side-by-side orientation of the cavities. Rather,malformations like inferosuperior ventricles, crisscross hearts, or dissociation between the ventricular morphologies and the distal truncal anatomy develop (i.e., arterioventricular dissociation, with L-loop great arteries and D-loop ventricles). Such atypical morphologies are most likely the result of atypical and complex intrinsic deformities of the bulboventricular loop (i.e., double looping).



The potential consequences of atypical cardiac looping are illustrated by this theoretical example of complex (double)looping. A) The upper looping is D, leading to right-sided aorta (normal); however, the lower looping is L, leading to a right ventricle on the left. B) The result is arterioventricular dissociation (D-great vessels, L-ventricles), which constitutes an exception to the "loop rule. "

Formation of the Left Ventricular Outflow Tract and the Right Ventricular Inflow Tract

- Shifting and Differential Growth The relationship between truncoconal outlets and ventricular structures becomes established simultaneously with another separate but equally important morphogenetic process: the acquisition of an inlet by the *bulbus cordis*.
- The early, looped cardiac tube has essentially a single inlet (the common atrioventricular canal), which conveys the venous blood to the primitive ventricle. (At this time, only systemic venous blood will be delivered, since there is no pulmonary circulation.) This primitive ventricle has no other outlet except a primitive ventricular septal defect (bulboventricular defect), through which blood is channeled into the bulbus cordis and from there into the common truncoconal tube .
- Later the atrioventricular (AV) canal grows by widening rightward, and the AV cushions acquire a direct relationship to the bulbus cordis. This relationship leads to the maturation of the bulbus cordis into the definitive right ventnicle .

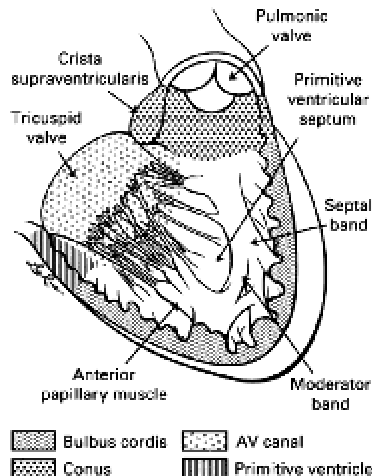


The acquisition by the early loop heart (A) of the final ventricular morphologies (B) requires a widening of the atrioventricular canal to the right, and a shifting of the conus to the left, with disappearance of the bulboventricular flange.These changes occur during the 5th and 6th weeks of human embryonic development. The arrows within these hearts represent blood flow patterns.

- At the same time, the arterial outlet undergoes a process of leftward shifting and differential growth that leads to the disappearance of the bulboventricular flange or spur ; the resorption of the caudal extreme; and leftward shifting of the conus,closer to the anterior AV canal cushion. This process is crucial; in its absence, a double-outlet right ventricular defect originates.

Developmental Implications.

- The early heart is characterized by a sequential arrangement of the cardiac segments (atria -> primitive ventricle -> bulbus cordis -> single truncoconal tube).
- In case of persistence of such a primitive arrangement, the most likely results would include a double-inlet left ventricle (or a common AV canal entering only the left ventricle) and a double-outlet right ventricle of the outlet chamber variety (without an AV valve or papillary muscles, and no inlet other than a bulboventricular communication).
- Normally formed right and left ventricles can be defined according to their embryologic components and intrinsic, anatomic features.
- The left ventricle is characterized by a single inlet, whereas the malformation called double-inlet "left" ventricle is the persistence of the embryologic primitive ventricle, a mitral valve, 2 papillary muscles, and an outflow tract. In embryologic terms, the *outlet portion of the left ventricle* (corresponding to the location of a supracristal ventricular septal defect) is a conal derivative,and not intrinsically a ventricular structure.
- Similarly, the right ventricle is characterized by a single inlet with a tricuspid valve, and 1 anterior papillary muscle connected to the ventricular septum by a moderator band. Part of the anatomic right ventricle (the outlet delineated by the crista supraventricularis)is a derivative of the embryologic conus .



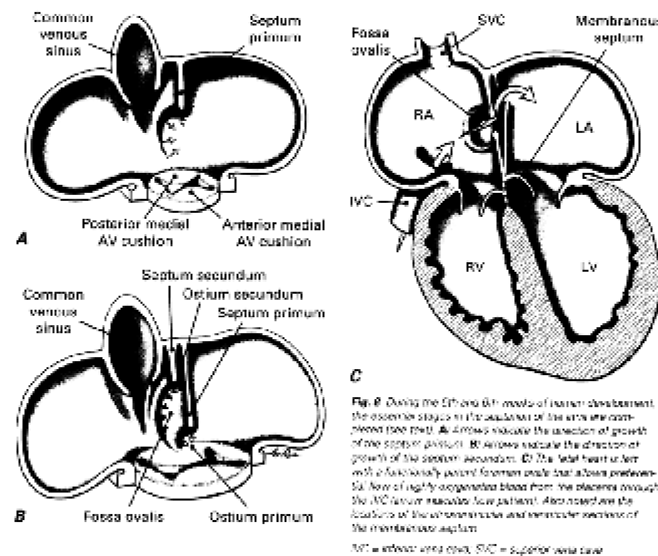
The definitive right ventricle is not made only from the bulbus cordis; it is also composed of tissues of the conus, the atrioventricular (AV) canal, and the primitive ventricle. Shown here is a cross-section of a right ventricle and its embryologic components.

- In anomalies like tricuspid or mitral atresia, the AV canal is normally aligned with the ventricular septum.
- Therefore, these anomalies are not related so much to the abnormal widening of the AV canal, as to the intrinsic defects in the differentiation of each AV canal subdivision.
- Such defects lead to atresia of either orifice, usually after normal closure of the inlet ventricular septum. Hence tricuspid atresia is not a form of double-inlet left ventricle, although, functionally,mixed venous blood enters only the left ventricle in both malformations. Tricuspid atresia can occur both with atresia of the annulus (in which the inlet of the right ventricle is also atretic) and with a normal (or only hypoplastic) tricuspid annulus with imperforated AV leaflets (in which case the inlet is variably but distinctly preserved).
- In the development of *double-outlet left ventricle*,the leftward shifting of the conus evidently *exceeds the normal shifting*, leading to the inclusion of most of the conal derivatives into the left ventricular out flow tract. In contrast, cases of double-outlet right ventricle suggest an *incomplete* left shifting of the conal derivatives. The occurrence of double-outlet right ventricle in conjunction with so many variants (for example, double side-by-side muscular coni,transposition, normally

crossed great vessels, common truncus, and tetralogy of Fallot) suggests that septation of the conus is a process independent of its leftward shifting.

Atrial Septation

- The normal, complete separation of the 2 atrial cavities is a complex process, involving ingrowths of tissue from different sources; duplication; fusion; and resorption.

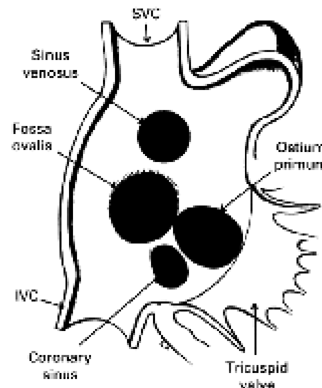


During the 5th and 6th weeks of human development,the essential stages in the septation of the atria are completed (see text). A) Arrows indicate the direction of growth of the septum primum. B) Arrows indicate the direction of growth of the septum secundum. C) The fetal heart is left with a functionally patent foramen ovale that allows preferential flow of highly oxygenated blood from the placenta through the IVC (arrow indicates flow pattern). Also noted are the locations of the atrioventricular and ventricular sections of the membranous septum.
IVC = inferior vena cava; SVC = superior vena cava

- First, a membranous structure appears on the posterosuperior aspect of the common primitive atrium medially to the entrance of the common venous sinus. This membrane, called the septum primum, grows caudally and anteriorly until it meets outgrowths of the medial AV cushions.
- The transient defect of the septum primum that is normally present in early embryos is called the ostium primum, which is closed when 3 structures converge and fuse: *the septum primum, and the anterior and the posterior medial AV cushions.*
- Before such convergence is completed, a new membrane,the septum secundum, appears just to the right of the septum primum, possibly as a duplication of the medial valve of the common sinus venosus. The septum secundum grows to cover the septum primum on the right side, with the exception of a round central area. This defect is called the fossa ovalis; *its floor, as seen from the right side of the atrial septum,*is composed of septum primum.
- *Normally, the septum primum also has or acquires a defect in its superoposterior section, called the ostium secundum.*Such a defect is not a true atrial septal defect (ASD), because it is covered on the right side by the septum secundum; however, it functions during fetal life as an obligatory escape valve, allowing highly oxygenated blood from the inferior vena cava (umbilical veins) to leak through the fossa ovalis and the ostium secundum into the left atrium.
- Only after birth do the septum primum and the septum secundum completely coalesce, so that the normal fetal patent *foramen ovale disappears, usually during the 1st 30 days of life.* In a limited percentage of otherwise normal hearts (5% to 20%), this defect remains potentially open (probe patent) for several years.

Developmental Implications.

§ Defects of the atrial septum typically occur at certain sites, in variable sizes :



The locations of the 4 most common types of atrial septal defects are illustrated in a right view of the atrial septum.IVC = inferior vena cava; SVC = superior vena cava

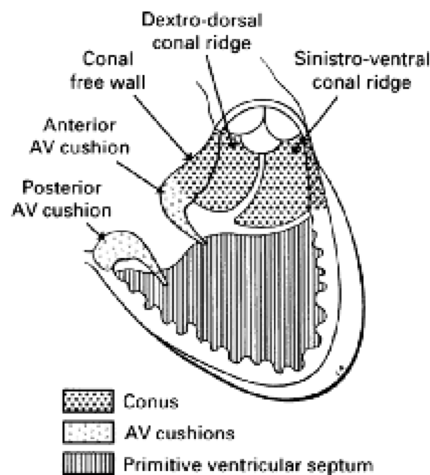
1) *Ostium primum ASD* is a defect of the caudal aspect of both the septum primum and the septum secundum, usually caused by or concomitant with a *lack of fusion of the 2 medial AV* cushions. This lack of fusion leads to the concurrent presence, in most ostium primum ASDs, of a *cleft in the mitral and tricuspid septal leaflets* or some type of AV canal defect.(See section on ventricular septal defects.)
2) *Fossa ovalis defect* (also-but improperlycalled secundum ASD) is the most common type of ASD and results from *resorption of the septum primum*,which normally provides the floor of the fossa ovalis. This defect should be distinguished from *persistence of the patent foramen ovale valve*, a virtually nonfunctional ASD (described above).
3) *Sinus venosus ASD* is located at the site of the normal defect in the septum primum called the ostium secundum, which becomes a permanent defect upon the lack of formation or the reabsorption of the adjacent septum secundum. *This defect is near the superior vena cava entrance into the right atrium and frequently occurs in conjunction with anomalous drainage of one or all the right pulmonary veins into the superior vena cava or the right atrium*. The reason for this association has not been proved, but it could be related to the jet effect directed at the upper atrial septum caused by a primarily abnormal flow from anomalous right pulmonary veins.
4) *Coronary sinus ASD* is a defect in the caudal posterior atrium, just above the normal site of drainage of the coronary sinus, which, in such cases, is missing its terminal section. The coronary veins drain directly into the left and right atria.
5) *Sinigle-atrium ASD* consists of a total absence of the atrial septum, frequently accompanied by clefts in the mitral and tricuspid septal leaflets or even a complete AV canal: evidence of the interdependence of the atrial septa and the AV cushions in normal development.

AV Canal and Ventricular Septation

§ Total closure of the ventricular septum is normally achlieved by the 45th day of developiment in the human embryo and depends on the convergence and fusion of 5 different components:

- the primitive ventricualr septum,
- the posterior and the anterior AV Cushlions,
- and the dextro-dorsal

Ø and the sinistro-ventral conal ridges .



Illustrated here are the 5 components of the normal ventricular septum that contribute to the final septal closure (at about the 7th or 8th week of development in the human embryo). The right ventricular side is shown.

§ The *primitive ventricular septum appears* shortly after the looping of the cardiac tube and defines the primitive ventricle as separate from the bulbus cordis. The upper edge of the primitive ventricular septum delineates the bulboventricular defect or primitive VSD, below the bulboventricular flange (or spur). In order to enable ventricular septal closing, the conus and AV canal must duplicate, and then align with the primitive ventricular septum; their components are essential to the definitive ventricular septum.

§ The final section of the ventricular septum to close is composed of permanently fibrous tissue,whereas the rest of the septum is composed of thick myocardial tissue. The normal site of the memibranous (fibrous) ventricular septum is just caudal and posterior to the crista supraventricularis, overriding the septal implantation of the tricuspid valve when viewed from the right ventricular side .

§ From the left ventricular side, the *membranous septum is located* below the aortic valve, between the right and the noncoronary cusps, in front of the bundle of His and above its anterior subdivision . Part of the fibrous septum then separates the left ventricular outflow tract from the right atrium,differentiating the implantation of the tricuspid valve (extending lower and anterior) from the mitral valve,which has a lower and posterior implantation into the ventricular septum . The anterior segment of the septal leaflet of the mitral valve constitutes the posterior wall of the left ventricular outlet.

Developmental Implications.

§ Lack of coalescence or primary failure of any component of the ventricular septum to form completely will result in a functional communication between the 2 ventricular cavities.

§ *Four types of ventricular septal defects (VSDs) are recognized* :

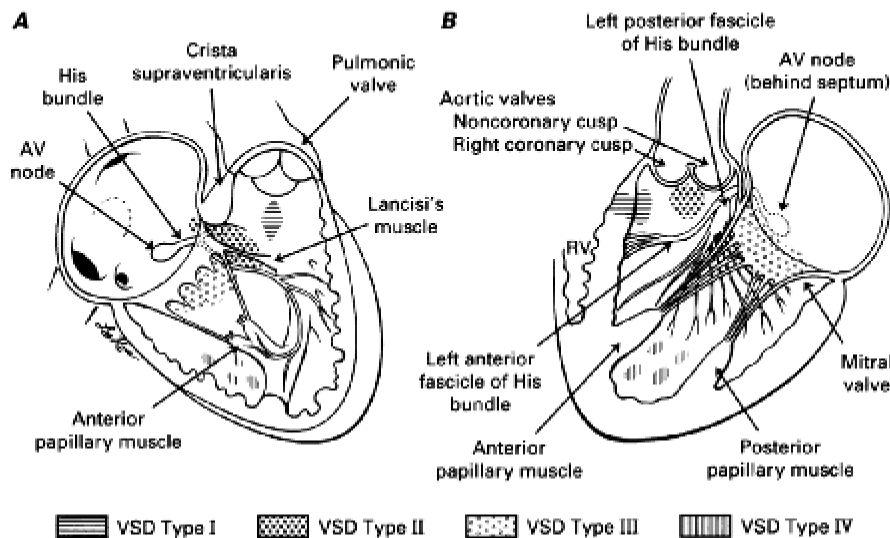
1) *Membranous or perimembranous septal defect* :

ü (VSD type II is caused by the failure of the membranous septum to form completely, and may occur because of an inadequacy of any of the 5 contributors.

ü This VSD is located in the left ventricular outflow tract, just beneath the noncoronary and right coronary cusps of the aorta.

ü Less frequently, this VSD consists of a left ventricular-to-right atrial communication (atrioventricular defect), typically located just above the anteromedial commissure of the tricuspid valve. From the right ventricular view, the membranous VSD is located just caudal

to the crista supraventricularis, anterior or posterior to the implantation of the *Lancisi's papillary muscle* of the tricuspid valve .



Typical locations of the different types of ventricular septal defects and the conduction system as seen from the right side of the septum (A) and the left side (B).

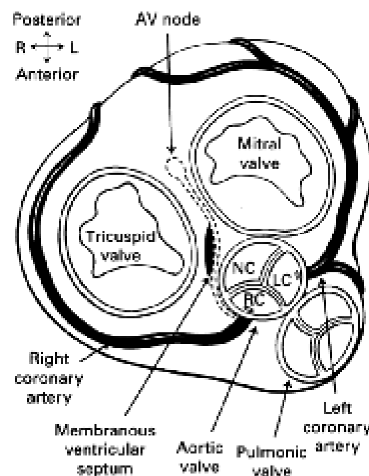
2) *Supracristal or conal septal defect* :
   - (VSD type I is located in the right ventricular outflow tract just below the pulmonary valve. From a left ventricular view, the defect is just below the right coronary cusp,which *frequently leads to the prolapse of that cusp into the defect* and consequent functional septal closure in diastole.Because of its close relationship with the supporting structure of the right aortic leaflet, this VSD is often complicated by aortic regurgitation. *It is interesting to note that the location of this defect indicates the midline of the conal septum and illustrates the contribution of this (conal) structure to the closing of the "ventricular" (anatomically defined)septum*.

3) *AV canal defects* (VSD type III)

- are the most complex VSDs; they involve not only a lack of separation of the ventricular cavities, but also some variable defects of AV valves. The variants range from the isolated cleft of the septal leaflets to a common AV valve with varying degrees of attachment to the ventricular septum and shortening of the height of the ventricular septum.
- Occasionally, subaortic stenosis is also associated with an AV canal defect.
- It is important to note that this VSD is caused by a failure of the uppermost component of the ventricular septum to develop below the septal leaflets of the AV valves, *most likely because of a lack in the medial AV cushions*.
- Similar to the type II VSD, the *membranous septum is never present* in these cases (these defects include a component of the type II VSD).
- A gooseneck appearance of the left ventricular outflow tract, indicated by left ventricular angiography, is characteristic of these VSDs. This configuration, as well as the presence of subaortic stenosis, may occur because the *septal leaflet (anterior half) of the mitral valve is usually implanted* into the *outflow tract of the left ventricle*, causing the outflow chamber to be hypoplastic and sometimes stenotic.
- The AV conduction system is also peculiar in these cases, because the bundle of His is displaced Incomplete inferoposteriorly, and its *anterior subdivision is elongated*. The left anterior fascicular block seen electrocardiographically in these cases is not a sign of real histologic or functional damage to the anterior fasicle, but simply of its increased length.

4) *Muscular septal defect* (VSD type IV)

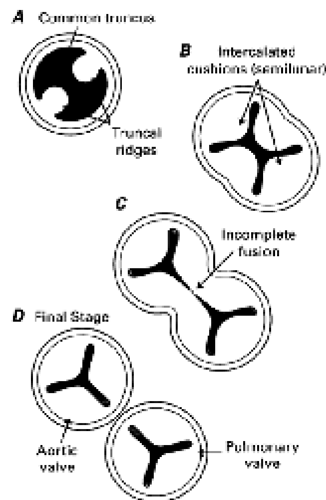
- is caused by 1 or multiple defects in the primitive ventricular septum.
- The location is unpredictable.
- These VSDs are usually small and difficult to recognize on surgical inspection from the trabeculated right ventricular aspect.
- In cases of tricuspid atresia, a peculiar and consistent type of muscular VSD occurs: a fissure-Aortic Pulmonary like, horizontal defect located inferiorly and posterioly to the membranous septum.
- Common ventricle is the case wherein little or no residuum of the ventricular septum exists, although the conal septum may be intact .This more severe lack of ventricular septation is probably due to the absence of both the primitive ventricular septum and the AV cushion's components.



The coronal section of the heart is illustrated, to show the location of the membranous ventricular septum and the course of the coronary arteries with respect to the cardiac valves.
LC = left coronary cusp; NC = noncoronary cusp; RC = right coronary cusp

Truncoconal Septation: The Formation of the Aortic and Pulmonary Trunks

- The earlier section on cardiac looping alluded to the basic and primitive relationship of the distal portions of the aorta and pulmonary artery (related to the development of the 4th and 6th aortic arches, respectively).
- The separation of the conus and truncus into 2 channels occurs simultaneously, but independently,at each level.
- The truncal septation is formed by *2 opposite (truncal) ridges*, which begin to develop from the area between the 4th and 6th aortic arches (the aortic sac) and grow caudally in a spiral fashion during normal development. *At their caudal extreme, the truncal ridges swell and are then called cushions*.
- They are complemented by similar intercalated cushions in the free wall of the common truncus .
- The further development of such “semilunar" cushions leads to the formation of the aortic and pulmonary valves.
- *Caudal to this level*, the *conal segment* (which is relatively longer in early embryonic hearts than in end-development hearts) presents a simultaneous process of duplication of the inner lumen.

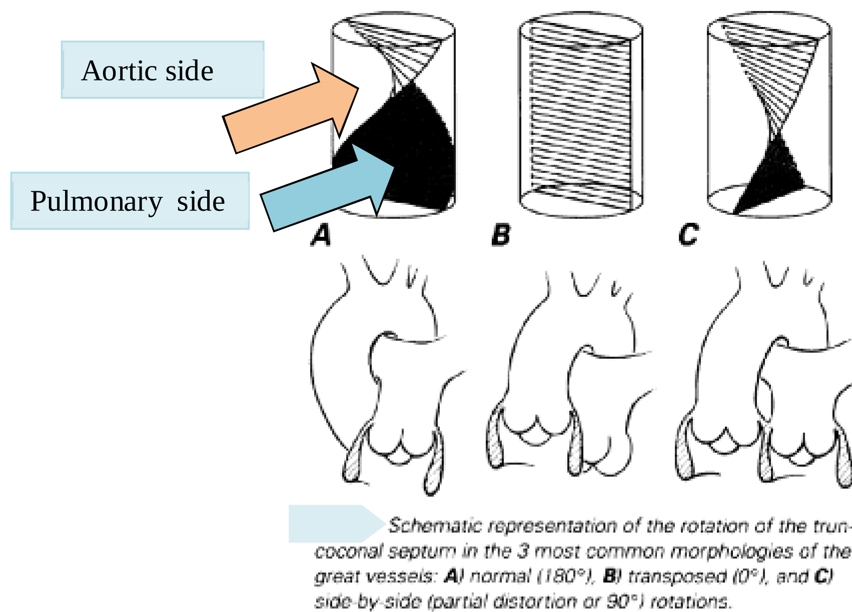


Formation of the pulmonary and aortic semilunar valves, by swellings (ridges) that appear at the caudal extreme of the truncus (completed by the 9th week in human embryos).

- This duplication is accomplished by the appearance of 2 opposite ridges (the dextrodorsal and sinistro-ventral conal ridges), which eventually fuse in the midline, as mentioned in the section on ventricular septation.
- It is interesting to recognize that in both normal and abnormal development, the truncal and conal septa correspond, even though they *develop by separate pro*cesses. Hemodynamic forces most likely lead to this alignment.
- In experimental embryology, the factors that lead to abnormal septation of the truncoconal segments have not been established. Recent investigations by Kirby and coworkers'" suggest the neural crest as a predominant influence. Early damage to the cephalad neural crest appears to result in the frequent occurrence of common truncus or transposition of the great vessels.

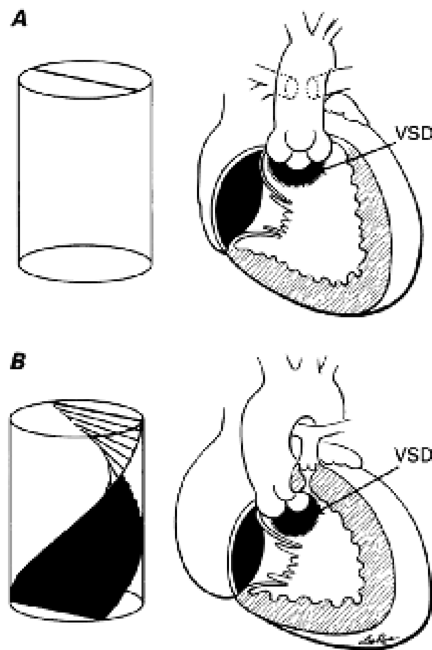
Developmental Implications.

- The arrangeimient of the great vessels is traditionally described as a reciprocal relationship in space (right to left, anterior to posterior), *so that normally crossed great vessels feature an anterior, muscular, pulmonary conus and a posterior, short, fibrous aortic outlet.*
- Since the 2 coniare connected to the 4th and 6th aortic arches,whose spatial relationship is established by cardiac looping, the normal resulting rotation of the truncoconal septum is as illustrated below.



Schematic representation of the rotation of the truncoconal septum in the 3 most common morphologies of the great vessels: **A)** normal (180°), **B)** transposed (0°), and **C)** side-by-side (partial distortion or 90°) rotations.

- In contrast, transposition of the great vessels features an aorta that is *parallel to the pulmonary artery,and positioned anteriorly and to the right of it* . The truncoconal septum is then straight.
- An *intermediate rotation* of the conotruncus leads to the *side-by-side arrangement* of the aortic and pulmonary valves that is seen frequently, but not exclusively, in *double-outlet right ventricle*.
- It is interesting to note that *anterior coni* are characterized by the higher location of the related semilunar valves and by a *muscular infundibulum*, whereas a *posterior conus* usually *features fibrous continuity* between the *related semilunar valve with the mitral and tricuspid valves*.
- Such intrinsic relationships of the great vessels could be defined independently of their connections with the ventricles.
- All 3 basic truncoconal morphologies can occur in *double-outlet right ventricles* (both great vessels arising mainly from the right ventricle); in *orthoposition* (1 vessel on eaclh side of the ventricular septum, as in the normal heart); or in *double-outlet left ventricle*.
- Recently, some investigators have found it more convenient to eliminate the description of the intrinsic relationship of the great vessels and describe connections instead. For example, the anatomy would be described as normal when the aorta arises from the left ventricle, and the pulmonary artery, from the right ventricle. The term transposition would be applied when the aorta arises from the right ventricle and the pulmonary artery from the left ventricle.
- According to this approach, further descriptions identify the infundibular morphologies. This system presents major difficulties because it does not identify the various and somewhat confusing spatial arrangements of the great vessels that occur in such anomalies as double-outlet right ventricle or left ventricle,or transposition of the great vessels. Only common agreement (among international societies) will settle these disagreements, which are only partially a matter of semantics.
- Malformations of the great vessels are not only characterized by the above-discussed variations in the *orientation* of the truncoconal septum, but by its *lack of development or uneven septation* of the conotruncus, leading to either pulmonary artery stenosis and atresia, or to aortic hypoplasia, stenosis,and atresia .



Truncoconal septation is totally absent in common truncus arteriosus (type 1I of Collett-Edwards illustrated in [Al) and unevenly displaced toward the pulmonary side in tetralogy of Fallot (B). In the latter anomaly, not only is the pulmonary valve stenotic, but also its annulus, the infundibulum,and the main trunk. The anteriorly displaced conal septum (normally completed) cannot align with the ventricular septum, leading to a ventricular septal defect (VSD)beneath an ectasic aorta.

- Common truncus arteriosus is, in its most complete form, the result of a lack of formation of the truncoconal septum in its entirety,which in turn leads to a single arterial trunk and semilunar valve, a conal septal defect, and the abgeneses are compared with normal development and are thoroughly understood.

Chapter 2

Segmental Diagnosis

Segmental Diagnosis

Cardiac Position
Visceroatrial Situs
A-V Valves, Ventricles
Conal Anatomy
The Great Arteries
Atrioventricular Alignments
Ventriculoarterial Alignments

- The segmental approach to cardiac diagnosis is based on diagnosing each segment of cardiac anatomy separately, based on its own characteristics instead of relying on associations. This system has been advocated by Van Proagh et al; we have followed it throughout this work.
- There are three cardiac segments - the atria, the ventricles and the great arteries. The atria connect to the ventricles via the AVcanal, and the ventricles connect to the great arteries via the conus arteriosus or infundibulum.
- Segmental diagnosis seeks to define each of these elements. Thus, cardiac position, visceroatrial situs, direction of ventricular looping, type of conal anatomy, the atrioventricular and ventriculoarterial alignments and associated malformations must all be defined individually.
- The echocardiographic approach to the diagnosis of complex congenital heart disease involves a segmental analysis of the heart. In this type of analysis, *one can think of the heart as being much like a house*. To describe a house completely, one must say where the rooms or chambers are located on each floor. For the "cardiac house," this description includes where each atrium is on the ground floor, where the ventricles are on the second story, and where each great artery is positioned at the top of the house. In addition, a complete description of the house should include the location of the staircases that connect the floors. For the cardiac house, this description includes atrioventricular and ventriculoarterial connections. *In this approach, if the atria are not correctly identified, the entire house comes tumbling down*.
- Thus, the approach to echocardiographic diagnosis of the patient with complex congenital heart disease *begins with a determination of the atrial situs* .
- *The word situs* refers to the topology or spatial position of the structure. *In atrial situs solitus*, the morphologic right atrium is on the right and the morphologic left atrium is on the left. *In situs inversus*, the morphologic right atrium is on the left and the morphologic left atrium is on the right. *In situs ambiguus*, the atria do not differentiate into right and left atria; instead, both atria can have features of *(1) a morphologic right atrium, a condition called asplenia, or (2) a morphologic left atrium, a condition called polysplenia.*
- The next step in the diagnosis of complex congenital heart disease is *determination of the bulboventricular loop* . This loop describes the locations of the ventricles. In a *d-loop (dextro loop), the morphologic right ventricle is on the right and the morphologic left ventricle is on the left. In an l-loop (levo loop), the morphologic right ventricle is on the left and the morphologic left ventricle is on the right.*

- *These definitions of d- and l-loop apply regardless of the atrial situs*. Thus,*concordant or normal connections* between the atria and ventricles (morphologic right atrium to morphologic right ventricle, morphologic left atrium to morphologic left ventricle) occur when there is situs solitus with a *d*-loop or situs inversus with an *l*-loop. Discordant or abnormal connections (morphologic right atrium to morphologic left ventricle, morphologic left atrium to morphologic right ventricle) occur when there is situs solitus with an *l*-loop or situs inversus with a *d*-loop.
- *In general, the convexity of the aorta points to the position of the right ventricle and thus helps indicate the bulboventricular loop.*
- *The definitive indicator* of the bulboventricular loop, however, is the *relative positioning of the ventricular inlets or atrioventricular valves*. Thus, in a *d*-loop the tricuspid valve is to the right of the mitral valve, and in an *l*-loop the tricuspid valve is to the left of the mitral valve.
- In most cardiac defects the inflow and trabecular portions of the right ventricle are on the same side relative to the components of the left ventricle, so that determination of the bulboventricular loop is straightforward (i.e., tricuspid valve to the right indicates *d*-loop, tricuspid valve to the left indicates *l*-loop). In some rare complex malformations discussed in detail later in this chapter, the inflow and trabecular portions of the right ventricle can be located on different sides of the left ventricular inflow (e.g., in certain forms of crisscross hearts). The spatial locations of the trabecular and outflow portions of the ventricles alone do not indicate the bulboventricular loop; their final spatial position is determined by the degree of apical rotation in the ninth week of gestation.
- *Normally, the cardiac apex pivots to the hemithorax opposite the bulboventricular loop*. In the early embryogenesis of the normal heart,the apex is oriented to the right (following initial rightward loop formation), but it subsequently rotates to the left as the primitive ventricular cavity develops into the left ventricle. Thus, in a normal *d*-loop the apex pivots to the left hemithorax; in a "normal" *l*-loop (i.e., one in the setting of situs inversus) the apex pivots to the right hemithorax. When the atrial situs and the loop are alignment concordant, apical pivoting is usually complete.
- Failure of complete apical pivoting is commonly associated with discordant atrioventricular connections. Partial pivoting in either concordant or discordant atrioventricular connections leads to a sagittally oriented ventricular septum and mesocardia. Rotational anomalies of the cardiac apex cause the ventricular septum and greater ventricular mass to be grossly displaced in space, whereas the inlet relationships of the ventricles are preserved. *Abnormal rotation can occur either in the frontal plane,along the longitudinal axis, or in both patterns.*
- Rotational anomalies of the cardiac apex also can cause the semilunar valves to be altered in their spatial relationships. An understanding of these apical rotational abnormalities and the resultant changes in the plane of the ventricular septum, the positioning of the greater ventricular mass, and the positioning of the great arteries is an essential component of the echocardiographic approach to segmental analysis of complex cardiac defects.
- If the echocardiographer does not understand the effects that rotational anomalies of the apex have on the standard imaging planes,imaging artifacts can be created easily and incorrect assessments made of the adequacy of chamber size.
- *In most congenital cardiac defects, there is harmony between the situs (topology) and alignment (connections) information which means that situs concordance nearly always predicts alignment concordance, and situs discordance nearly always predicts alignment discordance*. For example,situs solitus with an *l*-loop (discordant situs information) almost always predicts discordant atrioventricular connections or alignments (right atrium on the right connected to left ventricle on the right).
- The rare and *notable exception to this rule is the heart with crisscross* atrioventricular relations (discussed later). In this case, knowledge of the atrial situs and the bulboventricular loop might be wrongly predictive of the alignment or connections of these segments. In other words, there is disharmony between the situs and alignment information.

- *The final step* in the diagnosis of complex congenital heart disease is a *description of the great artery connections* . In normal or concordant connections, the pulmonary artery arises from the morphologic right ventricle and the aorta arises from the morphologic left ventricle.
- *Normally, the aortic valve is situated posterior and to the right of the pulmonic valve.*
- *Transposition is a discordant* ventriculoarterial connection in which the aorta arises from the morphologic right ventricle and the pulmonary artery arises from the morphologic left ventricle.
  - Classically, the old definition of transposition was based on spatial relations of the great arteries, that is, that transposition is present when the aortic valve and ascending aorta are anterior to the pulmonary valve and main pulmonary artery.
  - The more current definition of transposition proposed by Van Praagh et al and used in this chapter is based on the ventriculoarterial connections and not the spatial interrelationships.
  - The literal Latin root meaning of the word *transposition* is from *trans* (across)and *positio* (placement). Thus, the great arteries are literally "placed across"the ventricular septum, with the aorta arising from the morphologic right ventricle *and* the pulmonary artery arising from the morphologic left ventricle.
  - The old definition of transposition based on the anteroposterior relations of the great arteries has been largely abandoned because of the existence of transposition with a posterior aortic valve and an anterior pulmonic valve in 11% of autopsy-proved cases of transposition with dextrocardia.
  - Likewise, when the ventricles are severely rotated,normally connected great arteries can have an anterior aortic valve and a posterior pulmonic valve.
- *Other types of great artery connections* include
  - double-outlet right ventricle, double-outlet left ventricle, and single outlet from the heart.
  - Three common forms of single outlet from the heart include *truncus arteriosus*, *aortic atresia*, *and pulmonary atresia*. In Van Praagh's system of nomenclature, any great artery relationship that is neither normally crossed nor transposed is referred to as being *malposed*.Thus, for the heart with both great arteries arising from the right ventricle, with the aortic valve anterior and to the right of the pulmonic valve, the preferred nomenclature is double-outlet right ventricle connection with *d*-malposed great artery relationships (not double-outlet right ventricle *and* transposition, which would be two mutually exclusive types of connections).
- The echocardiographic examination is performed, and the interpretation is presented using a segmental approach , which requires complete definition of eight features of cardiac anatomy .
- Accurate morphology can be accomplished definitively only by imaging chamber septal structures. Next, other malformations (e.g., cardiac shunts, valve function) and physiology (biventricular function, chamber sizes,pressure estimates) are described.
- To ensure that no anatomy or physiology is left undescribed, it is helpful during both the performance and interpretation of the examination to imagine the course of a red blood cell traveling through the heart, beginning in the systemic veins and terminating in the systemic arteries.

SEGMENTAL APPROACH TO DEFINING CARDIAC ANATOMY BY ECHOCARDIOGRAPHY	
Anatomic Feature	*Diagnostic Possibilities*
1. Thoracoabdominal situs	Solitus Inversus Ambiguous
2.Cardiac position	Levocardia Mesocardia Dextrocardia
3. Atria	Solitus (S) (LA to the left of RA) Inversus (I) (LA to rhe right of RA) Ambiguous (A)bilateral right-and left-sidedness
4. *Atrioventricular connection*	Concordant Discordant Atresia Double inlet Common Straddling Crisscross
5. Ventricles	d-looped (D)(LV to the left of RV) l-looped (L) (LV to the right of RV)
6.Conus	Subpulmonic Subaortic Bilateral Absent or very deficient
7.Ventriculoarterial connection	Concordant Discordant Double outlet Single outlet
8.Great vessels	Solitus (S) Inversus (I) Transposed (D, L, A) Right and anterior (D)

A complete segmental description of the heart by echocardiography begins by defining these eight features, which include the three segments of the heart (3,5,8) and the two junctional segments (4,7).

a.Describing morphology:

The letters in parentheses following the diagnostic possibilities for the three cardiac segments are frequently used as abbreviated three-letter descriptors of these segments: The first initial describes atrial situs, the second describes ventricular topology, and the third describes the great vessel relationship. For example, (S, D, S) describes a normal heart, whereas (I, L, I) describes mirror image dextrocardia.

b.Describing pathology:

Following description of the morphology, intrinsic (e.g., pulmonary stenosis), myocardial (e.g., hypertrophic cardiomyopathy), septal (e.g., tetralogy of Fallot), orientation (e.g., crisscross heart), defect (e.g., ventricular septal defect or aortopulmonary window) pathology is described.

Cardiac Position

This refers to the spatial orientation of the heart in the thoracic cavity. The possibilities are:

Mesocardia: The heart is predominantly in the midline, with the apex pointing directly inferiorly.

Levocardia: The heart is in the normal position - predominantly in the left hemithorax, with the apex pointing leftwards and inferiorly.

Dextrocardia: *Dextrocardia* indicates that the heart lies in the right side of the chest. *There are three main categories of dextrocardia*.

- The first is termed *dextroposition* and refers to a condition in which the heart is pushed into the right chest by either a mass in the left chest (e.g., diaphragmatic hernia) or deficiencies in rightsided chest structures. The apex remains pointed to the left; atrial situs, ventricular topology, and great vessel relationships are normal; and cardiac pathology, if any, is not complex. *In the other two types of dextrocardia, termed dextroversion and mirror-image dextrocardia, the apex is pointed to the patient's right*.
- *Dextroversion* refers to a condition in which there is atrial situs solitus and ventricular inversion *atrioventricular discordance*)and associated pathology similar to that seen in congenitally corrected transposition of the great arteries.
- The final type of dextrocardia is called *mirror-image dextrocardia* with *atrial situs inversus*.
- There may be major and complex pathology associated with this type of dextrocardia. However, in situs inversus totalis (a condition in which all body organs are on the contralateral side of the body from which they usually reside), the heart may be completely normal.

- *Rotational Anomalies of the Cardiac Apex*

  Normal Heart
  - Apex displaced 30 to 60 degrees from a vertical direction in the frontal plane
  - Ventricular septum tilted so that its anterior edge is leftward and superior to its posterior edge

  Superoinferior Ventricles
  - Abnormal tilting of the apex in the frontal plane
  - Resultant horizontal ventricular septum and superior right ventricle

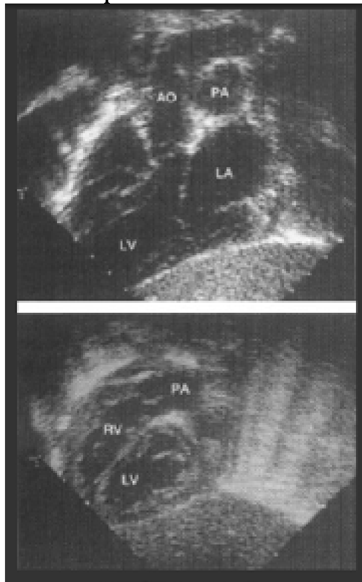
  Crisscross Ventricles
  - Abnormal apical rotation along the longitudinal axis

  Dextroversion
  - Lack of apical pivoting in situs solitus, d-loop
  - Resultant abnormal positioning of the apex to the right
  - Preservation of the relationship between the great arteries

Evaluation of the Patient with Dextrocardia

- The term *dextrocardia* simply indicates *that the heart is located primarily in the right chest and implies that one of three conditions is present* .
- *First*, dextrocardia can occur because the heart is displaced into the right chest, either because of a space-occupying mass in the left chest or because of absence of the normal lung volume filling the right chest. This form of dextrocardia is commonly called *dextroposition*.
- *Second*, dextrocardia can occur because of failure of pivoting of the cardiac apex to the left. This condition is known as *dextroversion* and is *frequently associated with atrioventricular discordance*.
- *Third*, dextrocardia can occur with abnormal atrial situs (i.e.,situs inversus or situs ambiguus). The most common condition in this category is *situs inversus totalis*, in which the heart is located in the mirrorimage position of normal.

- When a patient is referred with a diagnosis of dextrocardia,the echocardiographic examination is ***begun from the subcostal position*** rather than from the parasternal window, the routine starting position. From the subcostal four-chamber view, patients with dextroposition have the morphologic right atrium and right ventricle to the right of the morphologic left atrium and left ventricle. Usually, the alignment of the major axis of the heart is normal (pointed toward the left) or rotated slightly vertically;however, the entire heart is shifted to the right of midline or to the retrosternal area. In patients with dextroversion the morphologic right atrium is to the right of the morphologic left atrium; however, the major axis of the heart is aligned from the left shoulder toward the right hip. In this condition the cardiac apex is to the right of midline and the atria are usually in their normal positions or shifted slightly to the right .



Subcostal four-chamber views from a patient with dextroversion of the cardiac apex. Top, The left-sided ventricle has a smooth septal surface and is therefore the morphologic left ventricle (LV). The LV gives rise to a vessel that arches and is therefore the aorta (AO). Bottom, The plane of sound has been tilted far anteriorly. The right-sided ventricle has a prominent moderator band in its apical portion and is therefore a morphologic right ventricle (RV). The RV gives rise to a vessel that dives posteriorly and is therefore a pulmonary artery (PA). Other echocardiographic views showed that this patient had atrial situs solitus; therefore, the atrioventricular and ventriculoarterial connections are normal.The only abnormality in this heart is failure of pivoting of the cardiac apex to the left.LA, left atrium.

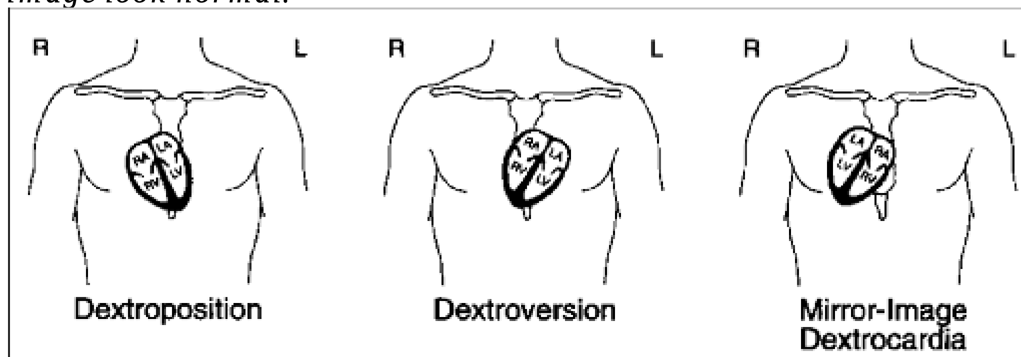
- In ***dextrocardia with atrial situs inversus***, the morphologic left atrium is to the right of the morphologic right atrium, and both atria are usually entirely to the right of the sternum . The cardiac apex is usually in the right fifth or sixth intercostal space at the anterior axillary line; therefore, the major axis of the heart is aligned between the left shoulder and right hip.In dextrocardia with atrial situs inversus, the parasternal long-axis view is obtained from the right second or third intercostal space with the plane of sound oriented in a mirror image of normal (from left shoulder to right hip).
- ***The parasternal long-axis view has only anteroposterior and superoinferior directions and does not display the right-to-left orientation of cardiac structures***. Thus, on the video monitor, cardiac structures appear to be oriented in a normal fashion, and only the examiner knows that the images were obtained in a mirror-image plane.
- The parasternal short-axis views are obtained from the same transducer location with the plane of sound also oriented in the mirror-image direction of normal (from right shoulder to

left hip). *Unlike the parasternal long-axis view, the parasternal short-axis view displays the right-to-left orientation of cardiac structures*. Thus, on the video monitor, in patients with dextrocardia and atrial situs inversus, the parasternal short-axis view appears to be a backward version of normal.

- It is important that the examiner not "correct" the image by using the leftright invert button. Inverting the images to make the views appear "normal"is contrary to the accepted guidelines for 2D image orientation and leads to confusion in understanding the spatial anatomy.
- In dextrocardia with situs inversus, the apical views are obtained with the transducer positioned in the right fifth or sixth intercostal space and with the plane of sound oriented in a mirror-image direction of normal. *Like the parasternal short-axis view,the apical four-chamber view in dextrocardia with situs inversus appears to be backward.*

ECHO TIPS:

- *When evaluating a patient with dextrocardia, the echocardiographer must maintain left/right conventions rather than attempting to make the image look familiar.*
- Specifically, the echocardiographer must remain true to the established echocardiographic convention that the *right* side of the screen/monitor in the parasternal short-axis, apical four-chamber, and subcostal coronal view is always the *left* side of the patient.
- This convention is maintained by the echocardiographer rotating the transducer so that the orientation mark is pointing to the patient's left in these imaging planes.
- *There will be a tendency (which must be resisted) for the echocardiographer to rotate the transducer to an unusual or atypical position to attempt to make the image look normal.*



Diagrammatic representation of the types of dextrocardia.

In dextroposition (in situs solitus),

- *the entire heart is shifted to the right chest, either because of a space-occupying mass in the left chest or because of absence of the normal lung volume filling the right chest.*
- *Usually, the alignment of the major axis of the heart is normal (pointed toward the left) or rotated slightly vertically;however, the entire heart is shifted to the right of midline or to the retrosternal area.*
- *The parasternal long- and short-axis views are obtained with the usual orientation of the plane of sound but with the transducer positioned just to the right of the sternum.*
- *The apical views are also obtained with the usual orientation of the plane of sound but with the transducer positioned just to the right of the lower sternal border.*

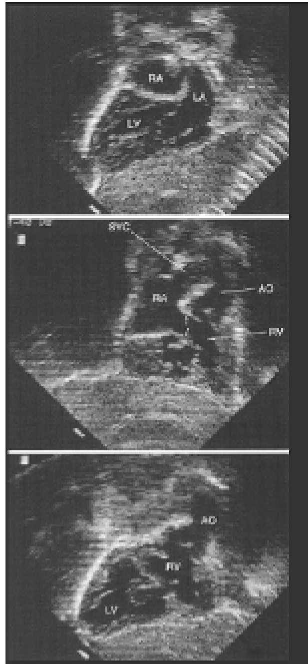
In dextroversion (in situs solitus),

- *there is failure of apical pivoting.*
- *The cardiac apex is to the right of midline and the atria are usually in their normal positions or shifted slightly to the right.*
- *The major axis of the heart is aligned from the left shoulder toward the right hip.*

- ü *The parasternal long-axis view is obtained with the plane of sound oriented in the mirror-image direction of normal.*
- ü *Because the atria are usually normally positioned and the great arteries arise normally from the ventricles, the parasternal short-axis view is obtained with a normal orientation of the plane of sound.* *In mirror-image dextrocardia or situs inversus totalis,*
- ü *the heart is located in the mirror-image position of normal.*
- ü *Both atria are usually entirely to the right of the sternum,*
- ü *and the cardiac apex is usually in the right fifth or sixth intercostal space at the anterior axillary line; hence, the major axis of the heart is aligned between the left shoulder and right hip.*
- ü *The parasternal long-axis view is obtained from the right second or third intercostal space with the plane of sound oriented in a mirror-image direction of normal (from left shoulder to right hip).*
- ü *The parasternal short-axis view is obtained from the same transducer location with the plane of sound also oriented in the mirror-image direction of normal (from right shoulder to left hip).*
- ü *The apical views are obtained with the transducer positioned in the right fifth or sixth intercostals space in the anterior axillary line and with the plane of sound oriented in a mirrorimage direction of normal. LA, left atrium; LV, left ventricle; RA, right atrium; RV,right ventricle.*

Crisscross Hearts

- § The term *crisscross heart* has been used to *describe the rare abnormality in which the systemic and pulmonary venous streams cross at the atrioventricular level without mixing.*
- § The right-sided atrium connects to the left-sided ventricle and the left-sided atrium connects to the right-sided ventricle.
- § This defect is believed to occur as a result of a differential rate of development of the right ventricular sinus and infundibulum. As a result, the ventricles appear to have rotated around their longitudinal axis (clockwise rotation when viewed from the apex in *d*-loop ventricles) without concomitant motion of the atria and atrioventricular valve annuli, producing actual crossing of the inflow tracts.
- § The defect can be found with concordant or discordant atrioventricular connections. *A ventricular septal defect is invariably present*, and discordant ventriculoarterial connections are common. Associated defects can be expected.
- § On 2D echocardiography, the diagnosis should be *suspected when a parallel arrangement of the atrioventricular valves and ventricular inflow regions cannot be found in the four-chamber views*. In the most posterior subcostal four-chamber view the left-sided atrium can be seen communicating by way of an atrioventricular valve to the right-sided ventricle. In the usual situation the left-sided atrium is a morphologic left atrium connected to a right-sided morphologic left ventricle. The left ventricle is posterior, inferior, and rightward. The posterior mitral valve is oriented from posterosuperior to anteroinferior .



Subcostal coronal views from a patient with a crisscross heart and dextrocardia. Top, The plane of sound is tilted far posteriorly. The pulmonary veins drain to the left-sided atrium, suggesting that this is a morphologic left atrium (LA). The morphologic LA is connected to a smoothwalled ventricle that has the anatomic features of a morphologic left ventricle (LV). The LV is located posteriorly, inferiorly, and rightward. The posterior mitral valve is oriented from posterosuperior to anteroinferior. Middle, The plane of sound has been tilted anteriorly so that the connections from the right-sided atrium to the left-sided ventricle can be seen. The right-sided atrium receives the drainage of the superior vena cava (SVC) and has other features suggesting that this chamber is a morphologic right atrium (RA). The anterior and superior tricuspid valve (arrows) is oriented from right to left and from posterior to anterior. A cross-section of the distal portion of the mitral valve leaflets can be seen inferior to the longitudinal section through the tricuspid valve leaflets. This view provides direct visualization of the crisscross arrangement of the atrioventricular valves. Bottom, Further tilting of the transducer shows that the left-sided ventricle has features of a morphologic right ventricle (RV) and gives rise to the leftward and anterior aorta (AO). Note that the morphologic RV is anterior, superior, and leftward.

- As the plane of sound is tilted further anteriorly, the connection from the rightsided atrium to the left-sided ventricle can be seen. In the usual situation the right-sided morphologic right atrium is connected to a morphologic right ventricle that is anterior, superior, and leftward.
- The anterior and superior tricuspid valve is oriented from right to left and from posterior to anterior. Although part of the tricuspid valve and right ventricular sinus extend to the left of the mitral valve, the annulus of the tricuspid valve is to the right of the annulus of the mitral valve. In this plane a distal portion of the mitral valve leaflets can often be seen in cross section inferior to the longitudinal section through the tricuspid leaflets. With even further tilting of the plane of sound anteriorly, the entire anterior ventricle and its outflow portion can be visualized. In the usual situation this anterior and superior ventricle is a morphologic right ventricle that gives rise to a transposed aorta.
- *Associated defects* are common in crisscross hearts. *Ventricular septal defects* are invariably present and usually occur in the inlet septum.
  Subvalvular and valvular *pulmonary stenosis* commonly occur. In most cases of crisscross heart, there is hypoplasia of the right ventricle. The degree of underdevelopment of the right ventricular sinus is directly related to the angle between the long axes of the atrioventricular valves and the degree of ventricular rotation. *Straddling mitral valve* is also commonly

encountered in crisscross hearts. In these cases the ventricles appear to have rotated through a greater angle, thus allowing alignment of the mitral valve and infundibulum.

- Although the ventricles in crisscross hearts are often arranged in a superiorinferior relationship, the terms "crisscross" heart and "superior-inferior ventricles" are not synonymous, and care should be taken not to confuse these entities on the echocardiographic examination. *Superior-inferior ventricles represent a distinct entity* characterized by a malpositioning of the ventricles with a horizontal ventricular septum.
- The defect is believed to occur as a result of an abnormal tilting of the cardiac apex in the frontal plane. In a study of 17 patients with superior-inferior ventricles,found crisscross relationships of the atrioventricular valves in 41% of the patients. Most patients (59%) did not have crisscross relationships.
- Crisscross heart is one of the few congenital defects in which knowledge of the atrioventricular connections or alignments is wrongly predictive of the ventricular spatial position. For example, knowledge of the atrial situs and the bulboventricular loop indicates the atrioventricular connections, which in turn usually correspond to the situs or position of the ventricles. Thus,the usual form of crisscross heart (right-sided morphologic right atrium to left-sided morphologic right ventricle to left-sided aorta) can be described
  as atrial situs solitus, *d*-loop, and *l*-transposition of the great arteries(Without further explanation, one would assume from this nomenclature that the atrioventricular alignments or connections are concordant and that the morphologic right ventricle is to the right of the morphologic left ventricle and this statement is not true).
- It is suggested, therefore, that in the rare instances in which there is disharmony between the situs and alignment information, both should be stated. The previously mentioned heart would then be referred to as atrial situs solitus, *d*-loop ventricles with crisscross connections, and *l*transposition of the great arteries.

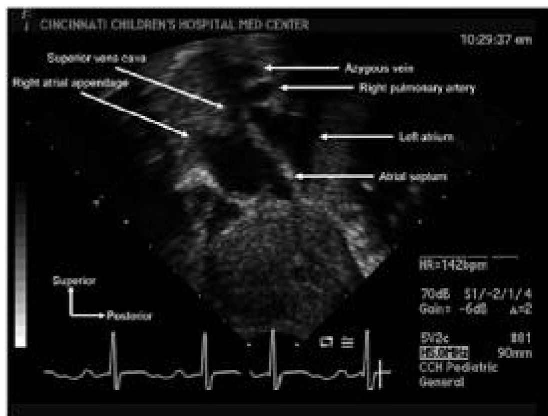
Visceroatrial situs

situs refers to the pattern of anatomic organization or arrangement of the *viscera and atria*.
The fact that an atrial chamber is on the patient's right or left or that it receives a particular venous structure does not allow a conclusion regarding atrial morphology. *Atrial situs can be deduced only by evaluation of the atrial appendages and the septal structures*.
The atrial appendages are consistently committed to their respective atria and are distinctly morphologically different.

- *The right atrial appendage* is short, fat, and broad based and best seen in the subcostal sagittal view .
- *The left atrial appendage* is long, thin, and narrow based and best seen in the parasternal short axis and apical four-chamber views .
- Although remaining committed to their respective atria, the atrial appendages may be *juxtaposed*, making their delineation more difficult.

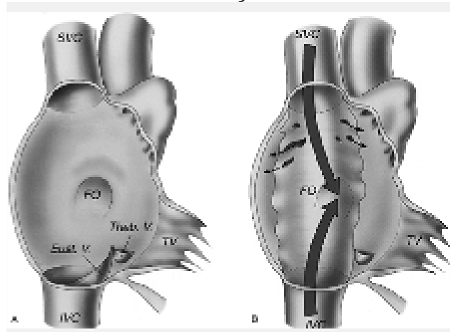


Parasternal short axis demonstrating the left atrial appendage, which is long and thin,distinguishing it from the right atrial appendage, which is short and broad. A, anterior; L, left; LA, left atrium; P, posterior; R, right; RA, right atrium.



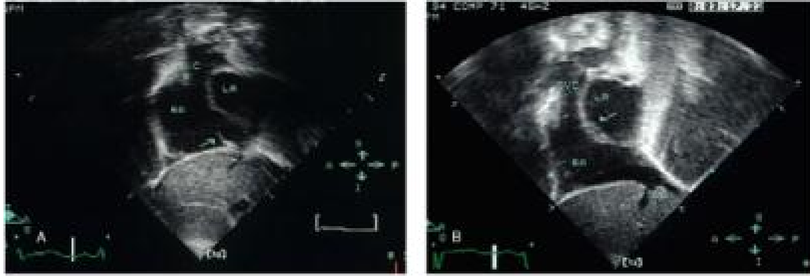
Subcostal sagittal view with rightward sweep demonstrating superior vena cava, left atrium,azygous vein, and right pulmonary artery. In this view the short and wide features of the right atrial appendage can be demonstrated.

- The unique *septal structures* of the *right atrium* (RA) are the embryonic valves(eustachian and Thebesian)seen in the subcostal coronal and sagittal views.



A: Diagram showing the normal appearance of the remnants of the right venous valve in the right atrium. The eustachian valve (Eust. V.) is seen at the entrance of the inferior vena cava (IVC) into the right atrium. The thebesian valve (Theb. V.) is seen at the right atrial orifice of the coronary sinus. B: Persistence of the embryonic right venous valve leads to diversion of the entire systemic venous return into the left atrium through the foramen ovale (FO). Multiple fenestrations of varying extent and size are often present (Chiari network) and allow some systemic venous blood to flow into the right ventricle through the tricuspid valve (TV). SVC, superior vena cava.

- The unique *left atrial septal* structure is the flap valve seen in the apical and subcostal coronal and sagittal views .



Subcostal sagittal views demonstrating eustachian valve (arrow in A) and flap valve (arrow in B). These unique septal structures are the most reliable method for defining the morphologic right and left atrium, respectively. A, anterior; I, inferior; IVC, inferior vena cava; LA, left atrium; P, posterior; RA, right atrium; S, superior; SVC, superior vena cava.

In heterotaxy syndrome with bilateral right- or left-sidedness and common atrium, the atrial septal structures will not be obvious and atrial situs will be indeterminate or ambiguous.

- *Situs solitus*: The liver is to the right, the stomach bubble to the left. The right atrium is to the right of the left atrium.
- *situs inversus*: The liver is to the left, the stomach bubble to the right. the right atrium is to the left of the left atrium,
- *Situs ambiguus*: The liver is midline; the stomach bubble variable in position. Sidedness of the atria cannot be definitively determined.This frequently occurs with visceroatrial heterotaxy.

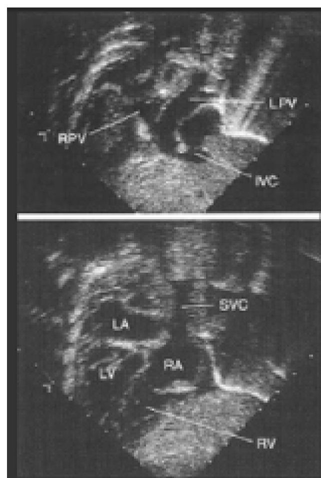
Echo Tips:

- *Best shown from subcostal views.*
- The position of the aorta and inferior vena cava shown from the transverse abdominal view from subcostal imaging) is a useful marker of situs(see appendex).Thus, the aorta is normally to the left and posterior of the inferior vena cava. In situs inversus, the aorta is to the right and posterior to the inferior vena cava.
- *Echocardiographic identifiers of the right atrium are the entrance of the coronary sinus and the inferior vena cava.*
- *The left atrium is identified by the flap valve of septum primum located on the left atrial aspect of the atrial septum.*
- Abdominal situs is best determined from a transverse view of the abdomen below the diaphragm .
- The position of the heart in the thoracic cavity is identified most easily from an apical four-chamber or subcostal coronal(long axis) view.
- The position of the transducer on the chest from where a standard apical four-chamber view is obtained defines the position of the cardiac apex.
- From the subcostal coronal view, the side of the cardiac apex can be ascertained by tipping the transducer slightly left or right .

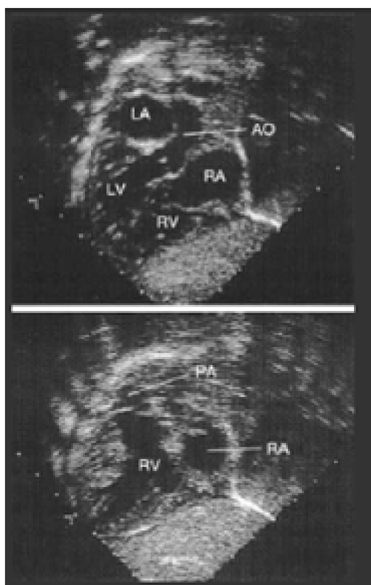




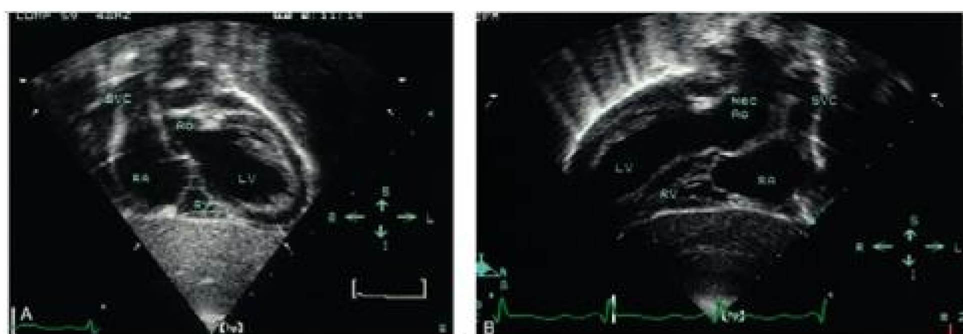
Subcostal transverse abdominal views in patient with abdominal situs solitus (A) and in a patient with abdominal situs inversus (B). With abdominal situs solitus, the liver is on the patient's right and the stomach is on the left. In abdominal situs inversus, the liver is on the left and the stomach is on the right. A, anterior; DAo, descending aorta; IVC, inferior vena cava; L, left; P, posterior; R, right.



Subcostal coronal views from a patient with atrial situs inversus, l-loop, and normal great vessels. This patient had total anomalous pulmonary venous return to the upper portion of the inferior vena cava (IVC) and a hypoplastic right lung. Top, The right and left pulmonary veins (RPV and LPV) can be seen connecting to a common pulmonary vein that drains below the diaphragm and connects to the upper IVC. Note that the RPVs are considerably smaller than the LPVs because of diminished drainage from the hypoplastic right lung. Bottom, The plane of sound has been tilted anteriorly to image the subcostal four-chamber view. Note that the heart is in the right chest with the apex to the right. The left-sided atrium receives the superior vena cava (SVC) and the IVC (top) and is therefore the morphologic right atrium (RA). The RA connects to a ventricle on the left that has an atrioventricular valve closer to the apex. These findings indicate that the left-sided ventricle is the morphologic right ventricle (RV) and that there is l-looping. LA, left atrium; LV, left ventricle.



Subcostal views from the same patients above . Top,The plane of sound has been tilted farther than that in Figure above in order to image the posterior great artery. Note that the morphologic left ventricle (LV) on the patient's right gives rise to a vessel that arches and is therefore the aorta (AO). Bottom, The plane of sound has been tilted even farther anteriorly to image the anterior great artery. Note that the morphologic right ventricle (RV) on the patient's left gives rise to a vessel that crosses from left to right anterior to the AO. This vessel is a normally connected pulmonary artery (PA). LA, left atrium;RA, right atrium.



Subcostal coronal views of a patient with levocardia (A) and dextrocardia (B). In the patient with dextrocardia (who also had an arterial switch operation for transposition of the great arteries so that the left ventricle (LV) now gives rise to the neo-aorta (Neo Ao), the apex of the heart is pointing to the patient's right. Ao, aorta; I, inferior; L, left; R, right; RA, right atrium; RV, right ventricle; S, superior; SVC,superior vena cava.

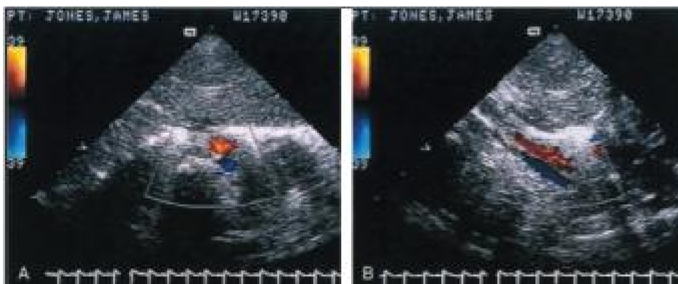
Situs Ambiguus

- The association of *splenic abnormalities* with complex cardiac defects and abdominal heterotaxy is well described.

  - *Asplenia*:
    - *right atrial isomerism*(Both Atria of RA morphology)
    - noting the presence of bilateral sinoatrial nodes and two morphologic right atrial appendages in these hearts.
    - the inferior vena cava and aorta tend to be on the same side of the spine, either to the right or to the left .

- total anomalous pulmonary venous return is nearly always present (in more than 80% of cases) and can be of any type. When the veins enter the cardiac atrium directly (rather than by way of a common pulmonary vein), they tend to drain to the smooth intercaval portion of the atria near the midline and are connected by a narrow confluence.
- *In asplenia syndrome, atrioventricular septal defects and single ventricle are common*. Most often, the single ventricle is not the classic double-inlet left ventricle; instead, a very rudimentary septum is present between the two ventricles.The great arteries are frequently transposed and there is a high incidence of severe pulmonary stenosis or atresia.

- *Polysplenia*:
  - *left atrial isomerism* (Both Atria of LA morphology) with two morphologic left atrial appendages.
  - drainage of the left-sided superior vena cava to the coronary sinus is encountered nearly always with polysplenia.
  - the inferior vena cava is frequently interrupted. In these cases, lower systemic venous return is by way of the *azygous or hemiazygous* vein, and the hepatic veins drain directly into one or both atria .
  - the pulmonary veins enter one atrium in a normal fashion in one third of cases. In over 50% of cases, however, the right veins enter the right-sided atrium and the left veins enter the left-sided atrium.
  - *Transposition of the great arteries and severe pulmonary stenosis are uncommon in polysplenia syndrome*.
  - In polysplenia syndrome, atrial septal defects,ventricular septal defects, and double-outlet right ventricle are often encountered.



Color Doppler examinations from a patient with polysplenia syndrome and an interrupted inferior vena cava. A, In the subcostal cross-sectional view, the descending aorta flow is seen in red (flow toward the transducer) because the transducer is situated in the abdomen and pointed slightly superiorly toward the patient's head. A large venous structure (blue flow area) is seen posterior and to the left of the descending aorta in the abdomen.
This posterior structure represents a large hemizygous vein through which the lower body systemic venous drainage returned to a left-sided superior vena cava. Flow in the hemizygous vein is seen in blue because the flow is directed away from the transducer and toward the patient's head. B, In the subcostal long-axis view, flow in the descending aorta is again seen in red, indicating flow down the aorta toward the transducer. Flow in the hemizygous vein (located posterior to the aorta) is seen in blue, indicating flow away from the transducer toward the heart.

A-V Valves and Ventricles

- The concept of ventricular looping is fundamental to the segmental approach to diagnosis.

- *In normal development the straight heart tube loops to the right (d-looping)*, thus bringing the right ventricle to lie to the right of, and anterior to the left ventricle. *Abnormal ventricular looping of the straight heart tube to the left (l-looping)brings the right ventricle to the left of the left ventricle.*
- The ventricles loop with the primordium for their corresponding A-V valves. *Thus, in d-looping, the right-sided A-V valve and ventricle are the morphologic tricuspid valve and right ventricle respectively.* In contrast, with ventricular l-looping the right-sided A-V valve and ventricle are the morphologic mitral valve and left ventricle respectively.
- The direction of ventricular looping can be surmised using the "chirality" "handedness"principle which holds that if the right hand can be positioned in the right ventricle with the palm on the septal surface and the thumb in the inflow and the remaining fingers in the outflow, then the ventricles are d-looped. If this maneuver can be performed with the left hand then the ventricles are l-looped.

Echo Tips:

- Best shown from subcostal and apical views.
- The *septal surfaces* of the two ventricles should be compared to decide which one is *more trabeculated*,which would suggest right ventricular morphology.
- *The offset of the A-V valves is an excellent echocardiographic marker of the direction of ventricular looping inpatients who do not have A-V canal defect*.
- Patients with d-loop ventricles have a normal offset, consisting of apical displacement of the septal leaflet of the right AV (tricuspid) valve relative to the left A-V (mitral) valve.
- The situation is reversed with L-loop ventricles, since the mitral valve is the right and the tricuspid valve to the left. Thus, with l-loop ventricles, there is apical displacement of the septal leaflet of the left AV(tricuspid) valve relative to the right AV (mitral) valve.
- The characteristics of the A-V valves are easily shown echocardiographically, and are useful indicators of the direction of ventricular looping.
- Thus, the *tricuspid valve* has chordal *attachments to the ventricular septum*; it opens towards the septum and has been described as septophilic.
- In contrast, the *mitral valve* has *chordal attachments with the free wall* and not to the ventricular septum; it opens towards the ventricular free wall and not towards the septum, and has been described as *septophobic*.
- *Thus, with d-loop ventricles, both A-V valves open towards the patient's left; with l-loop ventricles, both A-V valves open towards the patient's right.*

Conal Anatomy

- The term refers to the *structure of the ventricular outflow tracts* and, therefore, the orientation of the great arteries.
- *Normal*: conal (infundibular)) anatomy is characterized by a *subpulmonary conus* and absence of a subaortic conus. As a result, the pulmonary valve is anterior, superior and leftwards of the aortic valve; there is fibrous continuity between the mitral, aortic and tricuspid valves.
- *Subaortic conus*: This is characterized by persistence of the subaortic couns and absence of the subpulmonary conus. As a result, the aortic valve is anterior and either to the left. or to the right of the pulmonary valve. There is fibrous continuity between the mitral, pulmonary and tricuspid valves. T Ids type of conal anatomy is typically seen with d-transposition (if the great arteries.

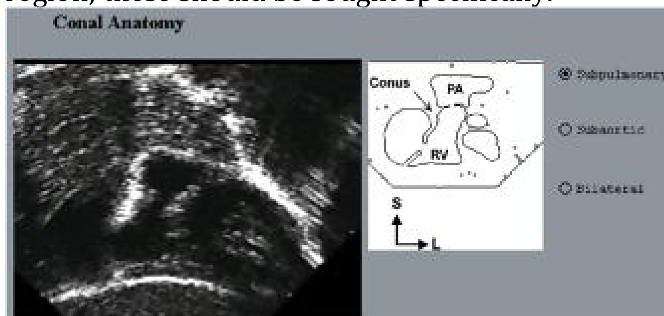
- F *Bilateral conus*: Conal tissue is present beneath the aortic as well as the pulmonary valves. This type of conal anatomy is typically seen with double outlet right ventricle.
- F *Bilaterally absent conus*: There is no conal tissue beneath either semilunar valve.Both A-V valves are in continuity with both semilunar valves. This type of conal anatomy is typically seen with double outlet left ventricle.

Echo Tips:

- Ø Best shown from subcostal long and short axis views.

Echocardiography should include evaluation for presence or absence of conal tissue beneath each semilunar valve.

- † If conal tissue is present, it should be examined for any deviation into the outflow tract lumen, and any resultant obstruction.Hypertrophy or hypoplasia of the conal septum should be sought.
- † Absence of conal tissue beneath a semilunar valve should prompt a search for other forms of outflow tract obstruction may exist, including A-V valve tissue billowing into the subvalvular region; these should be sought specifically.



Subpulonory conus(normal conal anatoy)
Subcostal long axis sweep
1.Pulmonary valve is located anteriorly and to left of aortic valve.
2. Pulmonary valve is separated from the tricuspid valve and right ventricular inflow by a prominent intervening tissue which comprises a subpulmonary conus ,this leads to the normal absences of the tricuspid to pulmonary valve continuity.
3.There is no such conal tissue beneath the aortic valve,as result there is direct continuity between the hinge points of the anterior mitral leaflets and coronary cusp of the aortic valve.

Great Arteries

The spatial orientation of the aortic and pulmonary valves is best characterized by describing the position of one valve relative to the other valve. This part of the segmental diagnosis assesses only the relative positions of the semilunar valves.

- ü *Normal* (Solitus): The aortic valve is located posteriorly and *rightwards* of the pulmonary valve {S,D,S}.(*S=atrial situs.D=ventricular looping.S=great artaries posion)*
- ü *Inversus*: The aortic valve is located *posteriorly* and *leftwards* of the pulmonary valve. This is typically seen in patients with situs inversus totalis and mirror-image dextrocardia(I,L,I}.
- ü *D-malposition*: The aortic valve is located *anteriorly* and *rightwards* of the pulmonary valve. This is typically seen inpatients; with transposition of the great. arteries with visceroatrial situs solitus and d-loop ventricles ;S,D,D }.
- ü *L-malposition*: The aortic valve is located *antetiorly* and *leftwards* of the pulmonary valve, This 's typically seen in patients with transposition of the great arteries with visceroatrial situs solitus and I-loop ventricles ;(S,LL).

- ü *A-malposition*: The aortic valve is directly *anterior* to the pulmonary valve. This is typically seen in patient with double outlet right ventrivle, and inpatients with transposition of the great arteries with visceroatrial situs solitus and d-loop ventricles {S,D,A}.

Eho Tips:

- Ø Best shown from subcostal and high parasternal short axis views.

The great arteries should be *identified by noting their course and branching pattern*; relying only on coronary artery anatomy to identify the great arteries maybe prone to error in the presence of anomalous coronary artery origin.

Atrioventricular Alignments

This aspect of segmental diagnosis *identifies which atrium opens into which ventricle*.

- † *A-V Concordance*: The right atrium opens into the right ventricle and the left atrium opens into the left ventricle.
- † *A-V Discordance*: The right atrium opens into the left ventricle and the left atrium opens into the right ventricle.

Ech Tips

- § Best shown from *apical and subcostal views*.
- § In order to identify A-V alignment correctly, the *visceroatrial situs and pattern of ventricular looping must first be determined*.
- § Thus, the echocardiographer should have identified which atrium is the right atrium and which ventricle is the right ventricle before any assessment of atrioventricular alignment can be made.

Venriculoarterial Alignments

This aspect of segmental diagnosis *identifies which ventricle opens into which great artery*.

- ü *Ventriculoarterial concordance*: The right ventricle opens into the pulmonary artery and the left ventricle opens into the aorta.
- ü *Ventriculoarterial discordance*: The right ventricle opens into the aorta and the left ventricle opens into the pulmonary artery.
- § In the next three forms of ventriculoarterial alignment, no decision can be made with regards to the concordance or discordance of ventriculoarterial alignment.

F For example, in a patient with double outlet right ventricle, the origin of the pulmonary artery from the right ventricle would be a concordant alignment; in the same patient, the aorta originates from the right ventricle, which would be a discordant alignment. Similarly, the term 'transposition', which refers to ventriculoarterial discordance, should *not be diagnosed* in a patient with double outlet right (or left) ventricle.

- ü Double outlet right ventricle: Both great arteries originate from one (right) ventricle.
- ü Double outlet left ventricle: Both great arteries originate from one (left) ventricle.
- ü Single great artery (truncus arteriosus): A single great artery originates from the heart.*Most commonly, a ventricular septal defect is associated, and the truncus overrides the ventricular septum*.

Echo Tips:

- Ø Best shown from *apical and subcostal views*.
- § In order to identify A-V alignment correctly, the visceroatrial situs and pattern of ventricular looping must first be determined.

- Thus, the echocardiographer should have identified which atrium is the right atrium and which ventricle is the right ventricle before any assessment of atrioventricular alignment can be made.

Chapter 3

Normal Structures

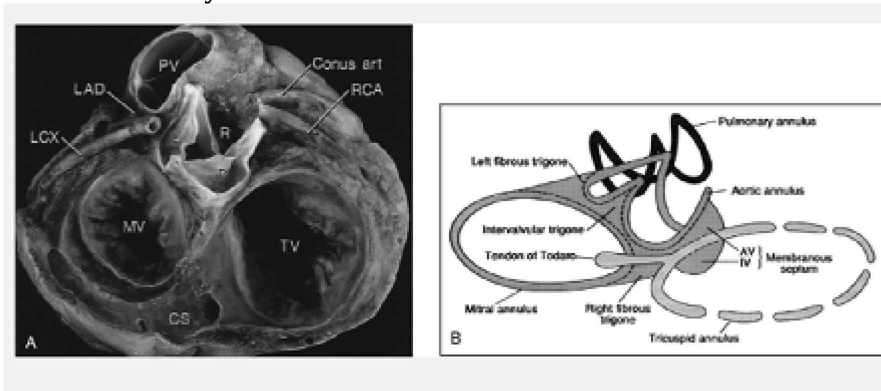
Normal Structures

- Thorough knowledge of normal cardiac anatomy and of the echocardiographic features of the normal heart are the essential foundation for understanding structural heart disease.
- This chapter describes the normal features of the cardiac chambers and great vessels.

Base of the Heart

- The cardiac base is defined by the plane of the atrioventricular groove (sulcus) and houses the four cardiac valves (Fig. below). It also contains the fibrous cardiac skeleton, whose purpose is to weld together the valvular annuli (annuluses), to fuse together but also electrically separate the atria and the ventricles, and to provide a firm foundation against which the ventricles can contract.
- The cardiac skeleton contains not only the four valve annuli but also their intervalvular fibrous attachments (the right, left, and intervalvular fibrous trigones and the conus ligament).
- The centrally located aortic valve forms the cornerstone of the cardiac skeleton, and its fibrous extensions anchor and support the other three valves.
- The *intervalvular fibrous trigone* is interposed between the left-posterior aortic commissure and the anterior mitral leaflet, and *the left and right fibrous trigones* project from each side and attach to the remainder of the anterior mitral leaflet. *Thus, the left, intervalvular, and right fibrous trigones provide the anatomic substrate for direct mitral-aortic valvular continuity.*
- The *membranous septum, in conjunction with the right fibrous trigone*, fuses the *right posterior aortic commissure to the anteroseptal tricuspid commissure*. Therefore, the right fibrous trigone (also known as the central fibrous body) welds together the aortic, mitral, and tricuspid valves and forms the largest and strongest component of the cardiac skeleton. Even in the setting of a membranous ventricular septal defect, this connection is maintained, so that the region of mitral-tricuspid continuity forms the posterior wall of the defect.
- Near the right-left aortic commissure is a diminutive connection between the aortic and pulmonary valves, the conus ligament (or ligament of Krehl).

- Thus, each aortic valve commissure is fused to one of the other three valves: *left-posterior commissure to mitral valve, right-posterior commissure to tricuspid valve, and right-left commissure to pulmonary valve.*
- Although schematic drawings of the cardiac base generally show the four valves in the same plane, they actually do not lie in the same plane or even in parallel planes. Because of the intertwining of the great arteries, the aortic and pulmonary valves are skewed 60 to 90 degrees as the valvular orifices are directed toward opposite shoulders. Moreover, the tricuspid and mitral valves are skewed 10 to 15 degrees, such that their annuluses approach one another at the membranous septum and diverge along the inferior wall as the coronary sinus is interposed between them. These angles may vary somewhat during the course of the cardiac cycle.



Base of the heart. A: The aortic valve is located centrally and abuts the other three cardiac valves. B: The cardiac skeleton, shown schematically, consists of four valvular annuli, three fibrous trigones, a membranous septum, and the tendon of Todaro.

Definition of Cardiac Chambers from the Two-Dimensional Echocardiogram

- Anderson et al proposed a rule of 50% for determining whether a cardiac chamber is a ventricle.
- This rule states *that a chamber is a ventricle if it receives 50% or more of an inlet. The inlet consists of the fibrous ring of the atrioventricular valve and need not always include a patent atrioventricular valve with wellformed valve leaflets.*

F For example, in hypoplastic left heart with aortic and mitral atresia, the fibrous ring of the mitral valve contains an imperforate membrane and is situated over the small left ventricle. Thus, this small leftsided chamber is a ventricle because it receives 100% of an inlet (even though there is no antegrade flow across the inlet).

- *A chamber need not have an outlet to be a ventricle*. Thus, the left ventricle in double-outlet right ventricle is a ventricle because it receives the mitral valve even though it does not have an outlet.
- The rule of 50% has also been used to *define the ventriculoarterial connections*. Thus, *if 50% or more of a great artery arises above a chamber, the great artery is defined as being connected to that chamber.*
- Application of the rule of 50% requires definitions for chambers that are not ventricles. According to the original descriptions,*rudimentary chambers* are chambers that receive less than 50% of an inlet and therefore do not qualify to be ventricles. There are two types of rudimentary chambers.

- ü An *outlet chamber* is one that has less than 50% of an inlet but 50% or more of an outlet or great artery.
- ü A *trabecular pouch* is a chamber that has less than 50% of an inlet and less than 50% of an outlet.

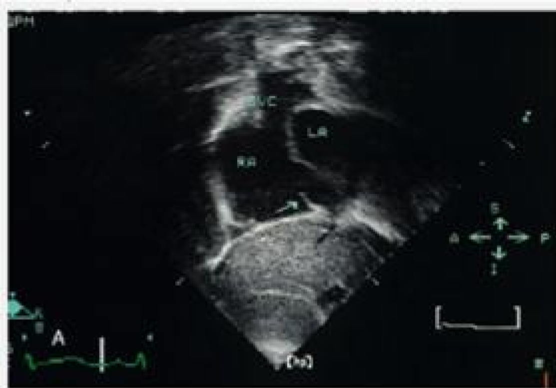
§ More recently, chick embryo studies by de la Cruz et al have shown that the trabeculated portions of the ventricles are the original developmental components. The inlet and outlet components form from the trabiculated component during and after looping; thus, the apical components are the oldest parts of the ventricles and form the basis for subsequent development. These and other observations form the basis for the belief by some investigators that chambers in the ventricular mass that possess a trabeculated portion of a ventricle should be considered ventricles regardless of whether they have an inlet or outlet component.Anderson has suggested that when such small chambers lack an inlet, they be called *rudimentary ventricles*.

Anatomic Landmarks on the Septal Surfaces

§ To diagnose complex congenital heart disease, one must know how cardiac chambers are identified on the 2D echocardiogram .

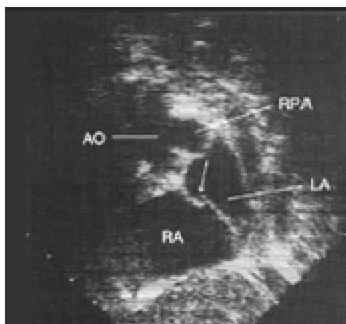
§ The cardiac chambers are largely defined by the *anatomic landmarks on their septal surfaces*.

§ *The morphologic right atrium* has a septal surface that *receives the tendinous insertion of the eustachian valve* and *has the limbus of the fossa ovalis*. *The eustachian valve* crosses the floor of the right atrium from the orifice of the inferior vena cava and inserts into the septum primum (the lower portion of the atrial septum adjacent to the atrioventricular valves). This tendinous insertion is along the lower border of the fossa ovalis and is called the *inferior limbic band* .



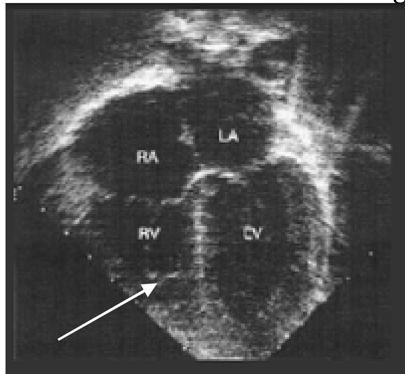
Subcostal sagittal views demonstrating eustachian valve (arrow in A)

§ The *left atrial septal surface* has the flap valve of the fossa ovalis. This is the *septum primum tissue* that covers the foramen ovale and seals it closed after birth .



Subcostal sagittal view from a normal patient. The flap valve (arrow) of the foramen ovale is seen on the atrial surface of the left atrium (LA). The flap valve of the foramen ovale is an anatomic marker of the morphologic LA. AO, aorta; RA, right atrium; RPA, right pulmonary artery.

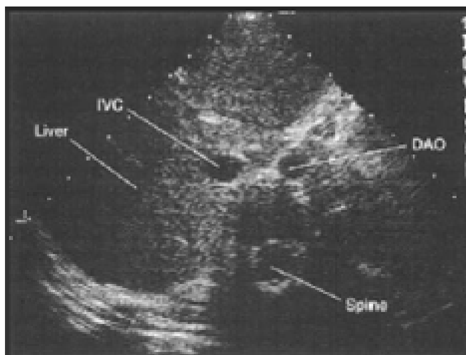
- The flap valve can be seen on the 2D echocardiogram protruding into the left atrium in the fetus when the foramen ovale is open; after birth, however, the flap valve is usually difficult to identify on the transthoracic 2D echocardiogram. With the use of high-frequency transesophageal imaging transducers, the flap valve can be imaged in a large proportion of patients. In cases in which the flap valve is tightly adherent to the left atrial septal surface and therefore cannot be visualized as a separate structure on the 2D echocardiogram, other methods for identification of the left atrium must be used.
- The *morphologic right ventricle* is the chamber whose septal surface has prominent muscle bundles crossing from the septum to the parietal free wall .



Apical four-chamber view from a normal subject. The morphologic right ventricle (RV) has prominent muscle bundles traversing from the septal surface to the parietal free wall and an atrioventricular valve closer to the cardiac apex(arrow). The morphologic left ventricle (LV) has a smooth septal surface and an atrioventricular valve farther from the cardiac apex. Note the pulmonary veins draining to the morphologic left atrium (LA). RA, right atrium.

- The largest of the septoparietal muscle bundles is the moderator band. In addition, the septal surface of the right ventricle receives chordal insertions from the tricuspid valve septal leaflet.
- *The morphologic left ventricle* is the chamber whose septal surface is smooth. There are no septoparietal free wall muscle bundles and the *mitral valve normally has no chordal insertions into the septum* .
- Another useful anatomic feature for identifying the ventricles is that the *atrioventricular valve always belongs to the appropriate ventricle*. Thus, the tricuspid valve is always found in the morphologic right ventricle and the mitral valve is always found in the morphologic left ventricle. The tricuspid valve is closer to the cardiac apex , has three leaflets, and has *chordal insertions* into the ventricular septum. The mitral valve is farther from the cardiac apex, is a fish-mouth bicuspid valve, and has chordal insertions only into two papillary muscles in the left ventricle .
- Systemic and pulmonary venous return can help identify the atria. *The pulmonary veins* usually drain to the morphologic left atrium; however, this is not a constant feature of the left atrium, because the pulmonary veins can drain anomalously. If three or more pulmonary veins drain by separate orifices to a chamber and there is no evidence of a pulmonary venous confluence, that chamber is most likely a morphologic left atrium. *The inferior vena cava* usually drains to the morphologic right atrium. This relationship is constant in most cases except in patients with situs ambiguous (discussed later). *The superior vena cava* usually drains to the morphologic right atrium; however, this relationship is not constant, as it can drain to either or both atria.

- The morphology of the *atrial appendages* can help identify the atria.The *right atrial appendage* is short and stout, resembling "Snoopy's" nose, and the *left atrial appendage* is long and finger-like, resembling "Snoopy's" ear .
- Also, the *abdominal situs* may provide helpful information for determining the atrial situs. For example, ***in most patients with atrial situs solitus, there is also abdominal situs solitus***. Thus, subcostal views of the abdomen show that the inferior vena cava is to the right of the spine, the descending aorta is to the left of the spine, the stomach bubble is on the left,and the liver is on the right . ***Likewise, in most patients with atrial situs inversus, there is also abdominal situs inversus***. Subcostal views of the abdomen show that the inferior vena cava is usually to the left of the spine and the descending aorta is usually to the right. The stomach bubble is on the right and the liver is on the left. In atrial situs ambiguus the liver may be to the right, to the left, or transverse. The stomach bubble can be on either side or in the midline. Several types of anomalies of systemic venous drainage often are present and suggest the diagnosis of situs ambiguus.



Subcostal short-axis view from a patient with atrial and abdominal situs solitus. Note that the liver is on the patient's right. The inferior vena cava (IVC) is to the right of the spine and the descending aorta (DAO) is to the left of the spine.

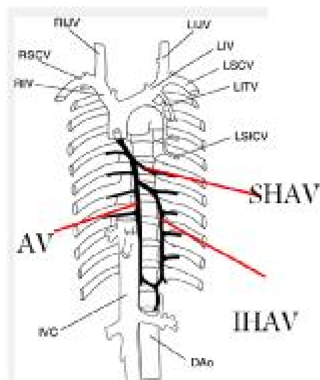
- ***When the atrial and abdominal situs are discordant (atrial situs solitus with abdominal situs inversus or vice versa), the incidence of severe, complex congenital heart disease is high.*** Defects with atrioventricular and ventriculoarterial discordance are frequent in this setting.

COMPARISON OF RIGHT-SIDED AND LEFT-SIDED ANATOMIC FEATURES OF CARDIAC SEGMENTS	
Right atrium	Left atrium
Limbus of fossa ovalis (limb of oval fossa)	Ostium secundum
Eustachian valve	The flap valve of the foramen ovale
Large pyramidal appendage	Small fingerlike appendage
Crista terminalis (terminal crest)	No crista terminalis
Pectinate muscles	No pectinate muscles
Receives venae cavae and coronary sinus[a]	Receives pulmonary veins[a]
Tricuspid valve	Mitral valve

Low septal annular attachment	High septal annular attachment
Septal cordal attachments	No septal cordal attachments
Triangular orifice (midleaflet level)	Elliptical orifice (midleaflet level)
Three leaflets and commissures	Two leaflets and commissures
Three papillary muscles	Two large papillary muscles
Empties into right ventricle	Empties into left ventricle
Right ventricle	**Left ventricle**
Tricuspid-pulmonary discontinuity	Mitral-aortic continuity
Muscular outflow tract	Muscular-valvular outflow tract
Septal and parietal bands	No septal or parietal band
Large apical trabeculations	Small apical trabeculations
Coarse septal surface	Smooth upper septal surface
Crescentic in cross sections[a]	Circular in cross section[a]
Thin free wall (3-5 mm)[a]	Thick free wall (12-15 mm)[a]
Receives tricuspid valve	Receives mitral valve
Pulmonary valve	**Aortic valve**
Empties into main pulmonary artery	Empties into ascending aorta
[a]Variable feature	

Systemic Veins and Right Atrium

- *The innominate(Brachiocaphalic) veins are identified in the suprasternal short-axis view.*
- Both innominate veins are identified and traced downstream as they join to form the *right superior vena cava (SVC)*, which, in turn, is followed to its entrance into the heart. The left innominate vein is traced upstream to identify a possible left SVC .

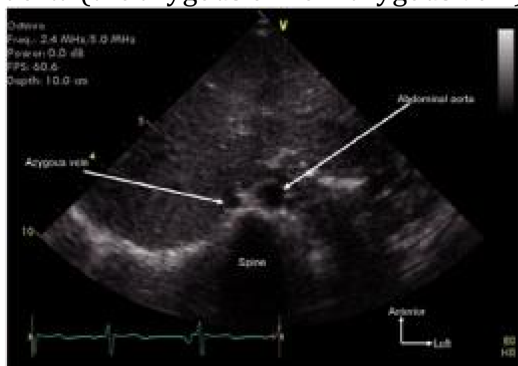


Schematic diagram of the azygos and hemiazygos veins (solid black) and their relations to the venae cavae. DAo, descending aorta; IVC, inferior vena cava; LIJV, left internal jugular vein; LITV, left internal thoracic vein; LIV, left innominate vein; LSCV, left subclavian vein; LSICV, left superior intercostal vein; RIJV, right internal jugular vein; RIV, right innominate vein; RSCV, right subclavian vein,AV=azigus vein SHAZV=superior hemi-azigus vein,IHAV= inferior hemi-azigus vein



Hybrid suprasternal notch oblique axis view of left superior vena cava (LSVC), which is partially fed by the hemiazygous vein. A small communicating vein (arrow) connecting the left and right superior vena cavae is also seen. A, anterior; I, inferior; P, posterior; S, superior.

- These maneuvers are particularly important in any patient undergoing surgical repair using cardiopulmonary bypass and in the newborn with single-ventricle physiology (who will undergo eventual cavopulmonary anastomosis).
- In patients with *unexplained cyanosis or systemic emboli or with absent innominate vein without dilated coronary sinus*, the presence of a left SVC draining directly into the left atrium (LA) should be investigated. This may require using agitated saline contrast .
- The inferior vena cava (IVC) is identified in the subcostal abdominal and cardiac views. Anyone performing cardiac catheterization needs to be alerted for interruption of the IVC, which is usually first apparent by imaging a large venous vessel adjacent or posterior to the aorta (the azygous or hemiazygous vein) in the short-axis abdominal view .



Subcostal transverse abdominal view demonstrating the features of interrupted inferior vena cava. There is no venous structure identified anterior to the aorta. Instead, there is a venous structure that is adjacent and slightly posterior to the aorta. The structure is also rightward of the aorta, indicating that it is an azygous continuation of interrupted inferior vena cava.

- *The RA and its appendage are best evaluated in the subcostal coronal and sagittal imaging planes*. Left juxtaposition of the RA appendage is usually evident in the parasternal long and short axes, where the wall of the appendage can be seen coursing perpendicular to the atrial septum.
- The entrance of the coronary sinus into the RA can be seen in the subcostal coronal and the apical four-chamber views. The size and possible unroofing of the coronary sinus can be assessed in a *posterior sweep* from the standard apical four-chamber and parasternal longaxis views.

The right atrium develops from :

- a primitive atrial component (the appendage),

- a venous component. (the entrance of the vena cavae and coronary sinus, derived from the embryonic sinus venosus)
- and an atrioventricular canal component (tricuspid valve, A-V canal septum).
- The exterior of the right atrium is characterized by abroad appendage with pectinate muscles.
- The right atrium receives the superior and inferior vena cavae and the coronary sinus.
- The superior vena cava enters the posterosuperior rightward aspect of the right atrium. Its entrance into the right atrium is *flanked by septum secundum to the left and crista terminalis to the right*.
- The inferior vena cava enters the posteroinferior rightward aspect of the right atrium Its entrance into the right atrium is guarded by the *Eustachian valve*.
- The coronary sinus enters the posterior inferior aspect of the right atrium *Immediately anterior to the entrance of the inferior vena cava*.
- On the septal surface of the right atrium, the space between septum primum below and septum secundum above is taken up by the foramen ovale.

Echo Tips:

- Best shown from apical, subcostal and parasternal short axis views.
- *The entrance of the inferior vena cava and the coronary sinus must be shown to confirm the identity of the right atrium.*

Right Ventricle

The right ventricle develops from the

- endocardial cushions (tricuspid valve, septum of the A-V canal),
- RV sinus (trabecular portion)
- and the conus(outflow tract).
- The external shape of the right ventricle is triangular. The right ventricle is tripartite, consisting of an *inflow*, *sinus and outflow tract*.
- The interior of the right ventricle has thick muscle bundles aid *numerous papillary muscles*. The papillary muscles attach to the septal surface; *the tricuspid valve is therefore septophilic, opening towards the septum*.
- The tricuspid valve forms part of right ventricular inflow, and is *unrelated* to the right ventricular outflow tract.
- The *right ventricular outflow tract* has an intricate architecture. It is bounded inferiorly by a *circumferential ring*, consisting of the septal band, moderator band and parietal band, and by the pulmonary valve above. The septal band runs along the septum and bifurcates superiorly.
- The conal septum fills the gap between the two divisions of the septal band. The parietal band is continuous with the conal septum and runs anteriorly onto the right ventricular free wall towards the apex where it is continuous with the moderator band at the base of the anterior papillary muscle.
  Septal band → conal septum → parietal band → moderator band → septal band
- The inferior rim of the infundibular outflow tract is completed by the moderator band extending from the parietal band to the inferior aspect of the septal band.

Echo Tips

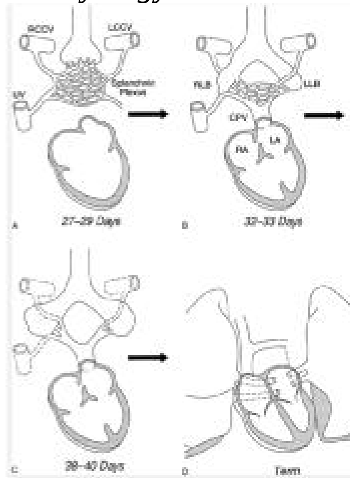
- Best shown from subcostal, parasternal short axis and apical views.
- *The right ventricle is defined by a trabeculated septal surface, tripartite nature of its interior and the septophilic tricuspid valve*.

- § Each of these should be defined to confirm the identity of the right ventricle.
- § *Enlarged right ventricle and should raise suspicion for a left to right atrial shunt, anomalous pulmonmary venous drainage, or severe tricuspid regurgitation.*

Pulmonary Veins and Left Atrium

- § Pulmonary veins:

Embryology:



Development of the pulmonary veins. A: At 27 to 29 days of gestation, the primordial lung buds are enmeshed by the vascular plexus of the foregut (the splanchnic plexus). At this stage, there is no direct connection to the heart. Instead, there are multiple connections to the umbilicovitelline and cardinal venous systems. A small evagination can be seen in the posterior wall of the left atrium to the left of the developing septum secundum. B: By the end of the first month of gestation, the common pulmonary vein establishes a connection between the pulmonary venous plexus and the sinoatrial portion of the heart. At this time, the connections between the pulmonary venous plexus and the splanchnic venous plexus are still patent. C: Next, the connections between the pulmonary venous plexus and the splanchnic venous plexus involute. D: The common pulmonary vein (CPV) incorporates into the left atrium so that the individual pulmonary veins connect separately and directly to the left atrium. LA, left atrium; LCCV, left common cardinal vein; LLB, left lung bud; RA, right atrium; RCCV, right common cardinal vein; RLB, right lung bud; UV, umbilical vein.

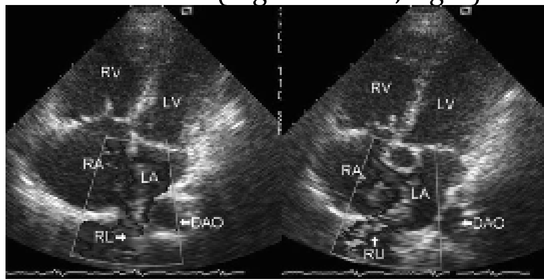
EMBRYOLOGIC CLASSIFICATION OF PULMONARY VENOUS ANOMALIES

- I. Normal absorption of the common pulmonary vein associated with defects that result in abnormal pulmonary venous drainage
  - A. Sinus venosus defect
  - B. Malposition of *septum primum*
- II. Atresia of the common pulmonary vein (early) while pulmonary-to-systemic venous connections are still present
  - A. Partial anomalous pulmonary venous connection
  - B. Total anomalous pulmonary venous connection
    - 1. Without pulmonary venous obstruction
    - 2. With pulmonary venous obstruction
- III. Atresia of the common pulmonary vein (late) after pulmonary-to-systemic venous connections are obliterated
  - A. Atresia of the common pulmonary vein
- IV. Stenosis of the common pulmonary vein
  - A. Cor triatriatum
- V. Abnormal absorption of the common pulmonary vein into the left atrium
  - A. Stenosis of the individual pulmonary veins
  - B. Abnormal number of pulmonary veins

Echo Tips

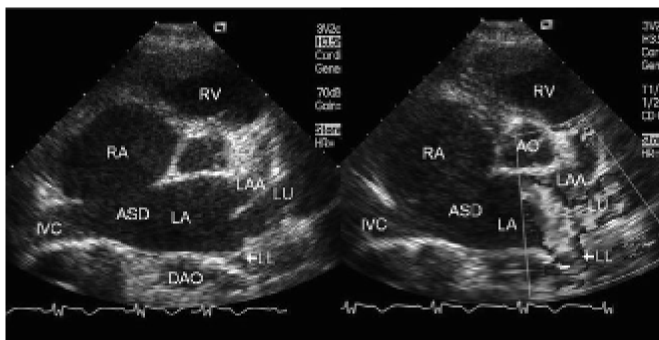
- § The proximal portion of the RLPV in the apical four-chamber view was seen draining into the LA almost perpendicular to its posterior aspect, and the blood flow of the vein was virtually parallel to ultrasound beam and atrial septum (Figure below,left).
- § Visualization of the RUPV made it necessary to tilt the transducer slightly anteriorly in the apical four-chamber view, when a portion of the left ventricular outflow tract could also be seen. Namely, lying between the apical fourand five-chamber views or near apical five-

chamber view was used, which entered, in a tilting position, the right postero-medial aspect of the LA (Figure below, right).



Doppler colour flow images of both right pulmonary veins. Left, the image of right lower pulmonary vein in the apical four-chamber view with colour flow mapping showing the vein draining into the LA almost perpendicular to its posterior aspect and virtually parallel to ultrasound beam and atrial septum. Right, the Doppler colour flow image of the right upper pulmonary vein in near apical five-chamber view indicates the vein in the right postero-medial aspect of the LA in a tilting position. DAO, descending aorta; LA, left atrium; LV, left ventricle; RA, right atrium; RL,right lower pulmonary vein; RU, right upper pulmonary vein; RV,right ventricle.

- The *parasternal short-axis view* at the level of the aorta and LA was the most ideal window to display the left upper pulmonary vein (LUPV). A length of the longitudinal section of the vein could be shown in all patients, which entered the LA through its lateral aspect in a slightly left anterior to right posterior direction (Figure below).
- It might be comparatively difficult to visualize the left lower pulmonary vein (LLPV)opening into the LA.



Two-dimensional and Doppler colour flow images of the left upper and lower pulmonary veins separated into the LA in the parasternal short-axis view in a patient with a mildly enlarged LA.Left, the left upper and lower pulmonary veins in the parasternal short-axis view showing separate opening into LA. Right, the Doppler colour flow image of both left pulmonary veins in this view. ASD, atrial septal defect; AO, aorta; IVC, inferior vena cava;LAA, left atrial appendage; other abbreviations as in Figure above.

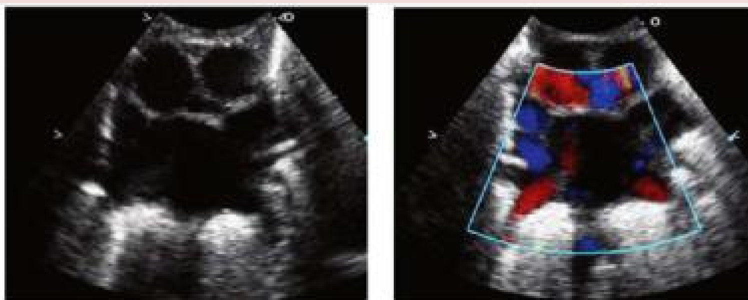
Contrast echocardiography		
	View	Course of entering the LA
RLPV	Apical four-chamber	Opening into the LA from its rear and almost parallel to the ultrasound beam
RUPV	Near apical five-chamber	Draining obliquely into the LA through its right posterior aspect
LLPV	Parasternal short axis	The vein entering LA through its lateral aspect with a slightly left posterior to right anterior direction

LUPV	Parasternal short axis	Draining into the LA in a slightly left anterior to right posterior direction

- The left atrium develops from :
  - a primitive atrial component (the appendage),
  - a pulmonary venous component (the common pulmonary vein
  - and an endocardial cushion component (mitral valve, A-V canal septum).
- The exterior of the left atrium is characterized by a thin, long finger-like appendage. The left atrium receives the pulmonary veins.
- The septal surface of the left atrium consists of septum *primum inferiorly* and *septum secundum superiorly*.

Echo Tips

- Best shown from subcostal and epical views.
- The entrance of the pulmonary veins into the left atrium posteriarly is shown from suprasternal short view (the so-called crab view).
- The pulmonary veins are identified most easily from the suprasternal short-axis plane.
- Alternatively, the pulmonary veins can be visualized from the parasternal short axis and apical four-chamber views.
- An extreme rightward sweep (tilt right from the SVC, RA, IVC views) in the subcostal sagittal plane consistently reveals the right upper pulmonary vein
- Color Doppler can aid in visualizing the individual pulmonary veins .
- Anomalous pulmonary venous pathways, the confluences from which they emanate, and the entrances of the pulmonary veins into the confluences should be investigated from the suprasternal short-axis view (supracardiac drainage), parasternal and apical views (cardiac drainage), and subcostal coronal and sagittal views (infracardiac drainage).
- Unlike systemic venous anomalies, in which color Doppler demonstrates flow coursing toward the heart, these anomalous pulmonary venous pathways will have color Doppler flow coursing away from the heart,often the first sign of which an echocardiographer becomes aware in this condition.



Normal pulmonary venous connection Suprasternal notch view of the pulmonary veins and left atrium with two-dimensional imaging (a) and color Doppler flow mapping (b) showing the "crabv view. Two left and two right pulmonary veins drain normally into the left atrium – the "legs" of the crab; the right superior vena cava and left atrial appendage are the "claws."

The flap of septum primum must be identifed; this confirms the identity of the left atrium. The left atrial appendage may be difficult to visualize erhocardiographically.



Subcostal sagittal view obtained by sweeping the transducer rightward from the systemic venous view consistently demonstrates the right upper pulmonary vein (arrow). A, anterior; I, inferior; LA,left atrium; P, posterior; RA, right atrium; S, superior.

Atrial Septum

- The atrial septum is examined in the *subcostal coronal and sagittal views*.
  - The size of an atrial septal defect,
  - as well as the degree of remaining atrial septal rim tissue for anchoring of a possible atrial septal defect (ASD)closure device
  - and the total atrial septal length for the maximum possible diameter for said device, is best performed in both these views to give information from two mutually orthogonal planes.
- Superior and inferior vena caval sinus venosus defects are best seen in the subcostal sagittal view(SAX). *The transducer should be swept gradually and deliberately posterior and rightward to investigate for possible associated partial anomalous pulmonary venous return*. Secundum atrial septal defects are best seen in the same imaging planes.
- The *ostium primum defect* is related to the crux of the heart seen in the *apical view*.
- The coronary sinus defect is visualized by sweeping the transducer posterior from the standard apical four-chamber view.

Atrioventricular Connection

Types

- The *septal structures of the atrioventricular valves* serve as the only consistent feature allowing morphologic diagnosis.
- The *tricuspid valve* has an intimate relationship with the ventricular septum, with multiple chordal attachments emanating from its septal leaflet, *seen best in the apical four-chamber view*.
- On the other hand, the *mitral valve* has no chordal attachments with the ventricular septum, and its entire attachments course to the left ventricular free wall.



Apical four-chamber view in a patient with ventricular inversion with congenitally corrected transposition of the great arteries {S, L, L}. The left-sided atrioventricular valve has attachments to the ventricular septum, whereas the right-sided valve does not, allowing diagnosis of a left-sided tricuspid valve and a right-sided mitral valve. In addition, the septal hinge point of the tricuspid valve is inferior to that of the mitral valve, which is another distinguishing feature of a tricuspid valve. There is a moderator band near the apex of the left-sided ventricle, further defining this ventricle as a morphologic right ventricle. I,inferior; LA, left atrium; RA, right atrium; S, superior.

- In addition, the *hinge point of the septal leaflet of the tricuspid valve* is inferior to the hinge point of the anterior leaflet of the mitral valve.
- Because each atrioventricular valve is associated with its appropriate ventricle (i.e., tricuspid valve with right ventricle, mitral valve with left ventricle), the atrioventricular connection can be determined as *concordant* (RA to right ventricle and LA to left ventricle) or *discordant* (RA to left ventricle and LA to right ventricle).
- *Atretic* atrioventricular connections are easily identified in the apical and subcostal views.
- The relationship of the atrioventricular valves to each other in double-inlet connections are explored in the parasternal views.

Mitral Valve

- The mitral valve is visualized in the parasternal, apical four-chamber, and subcostal coronal and sagittal views.
- The size of the *mitral annulus*, which is important in determining suitability for biventricular repair in cases of relative left-sided hypoplasia, should be performed in the apical four-chamber view.
- The *papillary muscles*, important to assess for repair of complete atrioventricular septal defect and for diagnosing parachute mitral valve, are best visualized in the parasternal short-axis and subcostal sagittal views.
- Mitral stenosis is assessed in the parasternal long-axis and the apical four-chamber views, where the degree of leaflet excursion can be seen clearly.
- Mitral valve prolapse is best identified in the parasternal long-axis and apical four-chamber views.
- Clefting of the mitral valve and double orifice mitral valve are usually seen in the parasternal short-axis sweep.

Tricuspid Valve

- The tricuspid valve is examined in the parasternal long-axis plane (sweeping right from the standard plane), the apical four-chamber view, and the subcostal coronal and sagittal views.
- The *septal leaflet* and its attachments to the interventricular septum are best seen in the apical four-chamber view. Also, in this view, the *posterior leaflet* (with a slight posterior sweep) or the *anterior leaflet* (with a slight anterior sweep) is seen on the lateral portion of the right ventricular wall. The anterior leaflet and its attachments to the conal papillary muscle (Lancisi) are best visualized in the subcostal coronal view sweeping anteriorly.
- In the evaluation of Ebstein's anomaly, the degree of atrialization of the right ventricle is assessed from the apical four-chamber view. From here, the septal attachments of the *septal leaflet* are appreciated. *The posterior mural leaflet* is seen with a slight posterior sweep from the apical four-chamber view. A portion of the *anterior mural leaflet* can be seen with an anterior sweep from the apical four-chamber view, but the subcostal coronal view is required to visualize the displacement of the anterior leaflet into the right ventricular outflow tract and the degree to which it obstructs it.

Ventricles

Ventricular Morphology

- Determination of the embryologic type of ventricular looping (d or l) first requires clear identification of ventricular morphology.
- The septal structures once again provide the definitive criteria for this evaluation. The *first criterion is the type of atrioventricular valve entering the ventricle* (see preceding section on Atrioventricular Connection, Type).
- The right ventricle also can be identified by its coarse, large, and extensive trabeculations along the septum and free wall. One of these trabeculations, the moderator band, is particularly prominent running transversely from free wall to septum in the inferior third of the right ventricular cavity in the apical view.

Left Ventricle

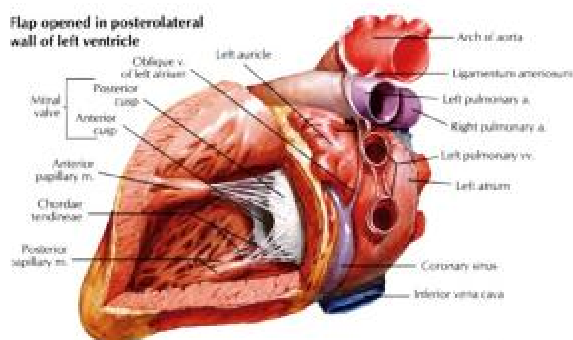
The left ventricle develops from:

- the endocardial cushions (mitral valve, septum of the A-V canal),
- the LV sinus (trabicular portion)
- and the conus (subaortic outflow tract).

- The external shape of the left ventricle is conical, like a football.
- The apical septal surface of the left ventricle has numerous fine trabeculae carneae, and is typically smooth superiorly.
- The mitral valve has two leaflets a large, sail-like anterior leaflet. and a smaller posterior leaflet.. There are usually two papillary muscles, which attach to the free wall; the mitral valve is therefore septophobic.
- The anterior mitral leaflet form the boundary between the left ventricular inflow and outflow tracts.
- Resorption of the subaortic conus leads to mitral-aortic fibrous Continuity.

Echo Tips

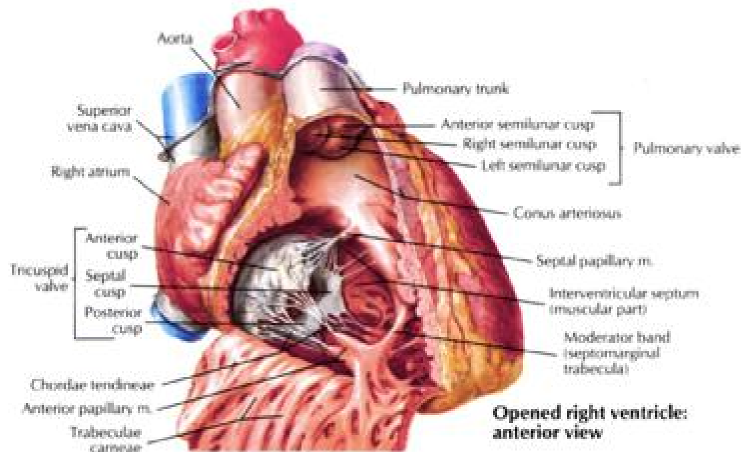
- Best shown from apical and parasternal short axis views.
- The size of the left ventricle, which is particularly important in evaluating atrioventricular septal defects and variants of hypoplastic left heart with relative left-sided hypoplasia, is evaluated in the apical four-chamber view.
- *The left ventricular outflow tract*, which is important to visualize for membranes and subvalvar stenosis, is seen by a slight anterior tilt of the transducer. Equally valuable are the parasternal long-axis view in which the left ventricular outflow tract is at a slightly shallower depth, improving imaging, or the subcostal coronal view.
- The *smooth septal surface*, *septophobic mitral valve* and *absence of an infundibulum* help characterize the left ventricle. These should be defined to confirm the identity of the left ventricle.



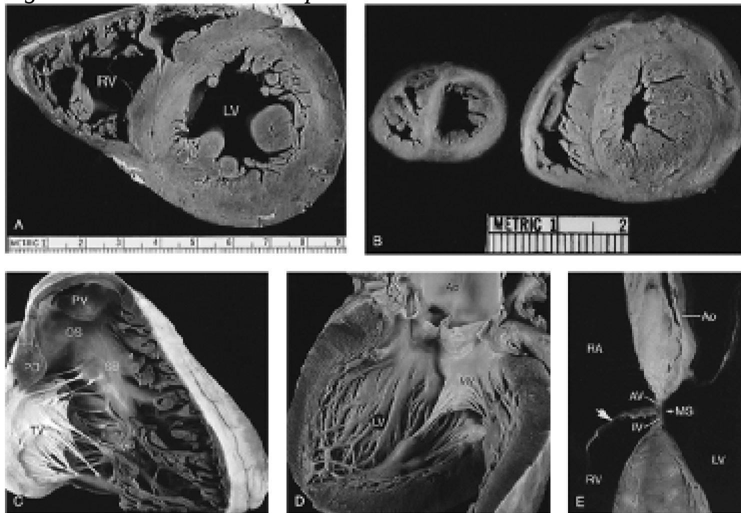
Left ventricle and the mitral valve

Right Ventricle

- The size of the right ventricle and its relative contribution to the ventricular apex in conditions such as complete atrioventricular septal defect and pulmonary atresia with intact ventricular septum are best assessed from the apical four-chamber view.
- Because the three portions of the right ventricle (inlet, trabecular, and conus[infundibulum]) do not lie in a single plane, visualization of the entire right ventricular cavity requires sweeping of the transducer through multiple planes in the subcostal coronal and sagittal views.



Right ventricle and the tricuspid valve



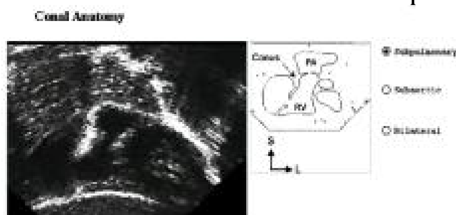
Comparison of right and left ventricles. A: In cross section, the right ventricle is crescent shaped, the left ventricle is circular, and the wall thickness of the left ventricle is three to four times that of the right ventricle. B: The fetal heart (to the left) exhibits prominent right ventricular hypertrophy, whereas by the age of 3 months, the infant's heart (to the right) shows regression of hypertrophy. C: The right ventricle receives a tricuspid valve, has prominent anteroapical trabeculations, and has a muscular outflow track that separates the tricuspid and pulmonary valves. The parietal (PB)and septal bands(SB) and the outlet septum(OS) form the crista supraventricularis (supraventricular crest). D: The left ventricle, in contrast, receives a mitral valve, has shallow apical trabeculations, and exhibits direct continuity between the mitral and aortic valves. E: The membranous septum is divided into atrioventricular and interventricular components by the septal tricuspid leaflet (arrow).

Ventricular Septum

- The ventricular septum is composed of two components: (a) the *membranous septum*, which is an extremely small (5 mm in diameter in the adult heart), and superior portion wedged between the tricuspid and aortic valves; and (b) the large *muscular septum*.
- The *membranous septum* (figure above)consists of two portions separated by the septal leaflet of the tricuspid valve: the pars atrioventricularis, where left ventricular to RA shunts occur, and the pars interventricularis, where ventricular septal defects are located.
- The *muscular septum* consists of three portions:
  - the *inlet portion*, which is inferior to the membranous septum and is between the atrioventricular valves;
  - the *trabecular portion*, which extends from the membranous septum to the apex;
  - and the *conal (or outlet or infundibular)* septum, immediately below the pulmonary valve.
- *Many ventricular septal defects typically occur along embryologic fusion lines* (e.g., a perimembranous outlet ventricular septal defect [VSD] is along the fusion line between the membranous and conal septa).
- *VSDs within the membranous* septum can be assessed in the parasternal long- and short-axis, apical five-chamber, and subcostal views. *In the basal short-axis view where the subpulmonary and subaortic regions can be imaged simultaneously, it can be determined whether an outlet ventricular septal defect is below the crista supraventricularis (infracristal outlet VSD) or above the crista (supracristal outlet VSD, also known as subpulmonic or doubly-committed VSDs)*. The membranous septum is seen well in the parasternal long-axis sweep from the standard view toward the tricuspid valve.
- In the apical view, the transducer should be swept anteriorly so that the left ventricular outflow tract and aorta are visualized.
- *VSDs within the conal septum* are assessed in the parasternal long axis sweeping left toward the pulmonary valve, in the basal short axis, and in the subcostal coronal and sagittal views.
- The *trabecular septum* is so large that defects within it need to be localized, preferably describing their position in two orthogonal planes and in relation to nearby landmarks. One classification system assigns the VSD as *anterior*, *midmuscular*, *apical*, *or posterior*.
  - The *anterior trabecular VSDs can be missed* if the echocardiographer is not interrogating consciously for them. Perhaps the best view is the parasternal long-axis view sweeping to the left.
  - The *midmuscular VSDs* can be seen in the standard parasternal long- and short-axis views and apical four-chamber views.
  - VSDs in the *posterior trabecular septum* are visualized best in the parasternal short-axis view or in the apical view swept posterior.
  - *Apical trabecular defects* are best seen in the apical four-chamber view inferior to the moderator band.
- *Inlet VSDs* are best visualized in the *short axis sweeping inferiorly* toward the atrioventricular valves and in the *standard apical four-chamber* view at the level of the atrioventricular valves. These are distinguished from atrioventricular septal defects by close echocardiographic inspection of the hinge points of the atrioventricular valve annuli, which remain normal (i.e.,mitral hinge point slightly superior to that of the tricuspid valve) with an inlet ventricular septal defect and at identical heights with an atrioventricular septal defect.

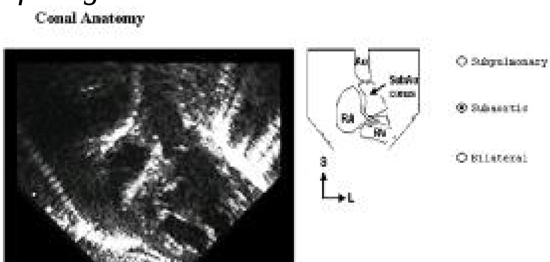
Conal Morphology

- *The conus (or infundibulum)* is the cavitary space formed by the muscular segment of the heart that *connects the ventricles with the great arteries* and *separates the atrioventricular and semilunar valves*.
- Abnormalities in conal development consist of variations in the presence, length, and diameters of the subpulmonary and subaortic conus. These variations can lead to (or be associated with) complex malformations, such as tetralogy of Fallot,interrupted aortic arch, transposition of the great arteries, and double-outlet right ventricle.
- *In the normal heart*, the conus is the nearly *vertical tubular outflow portion* of the right ventricle, which is separated from the nearly *horizontal right ventricular inflow portion* by distinct muscle bands. These muscle bands form a near-circular rim formed by the *parietal band anteriorly*, *the crista supraventricularis posteriorly,and the septal band medially and prohibit pulmonary valve to atrioventricular valve continuity*.
- *Subpulmonic :* The subpulmonary conus is best identified in the *subcostal views*. *Leftward anterior deviation* of the conal septum leading to a narrowed conus and subvalvar pulmonary stenosis in tetralogy of Fallot is evident in these views. These relationships can be demonstrated in the subcostal coronal and sagittal views. *Posterior deviation of the conal septum and ventricular septal defect results in left ventricular outflow tract obstruction and is associated with interrupted aortic arch*. The conal septum in this lesion is best assessed from the parasternal *long-axis view*.



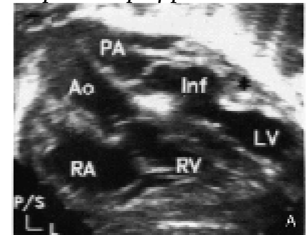
Subpulonory conus(normal conal anatoy)
Subcostal long axis sweep
1.Pulmonary valve is located anteriorly and to left of aortic valve.
2. Pulmonary valve is separated from the tricuspid valve and right ventricular inflow by a prominent intervening tissue which comprises a subpulmonary conus ,this leads to the normal absences of the tricuspid to pulmonary valve continuity.
3.There is no such conal tissue beneath the aortic valve,as result there is direct continuity between the hinge points of the anterior mitral leaflets and coronary cusp of the aortic valve

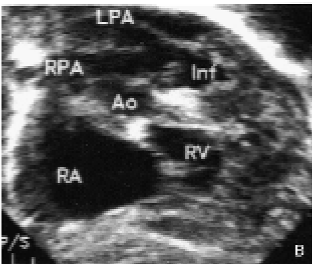
- *Subaortic:*Persistence of the subaortic conus and involution of the subpulmonic conus is the usual conal relationship in d-(or l-) transposition of the great arteries . The subaortic conus is evident on the subcostal coronal and sagittal views. Persistence of subaortic conus prohibits continuity of the aortic valve to either atrioventricular valve, and involution of the subpulmonary conus allows continuity between the pulmonary valve and both atrioventricular valves in transposition of the great arteries. *An extremely unusual variant of conal morphology is persistence a subpulmonic conus in the setting of d-transposition of the great arteries .*

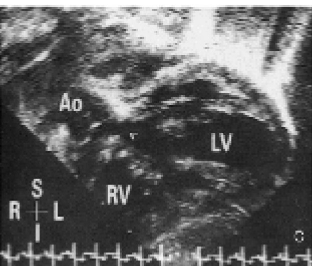


Subaortic conus

Subcostal long axis sweep(in patient with D-Transposion of graet artaries).
1.Aortic valve located anteriorly and right wards of pulmonary valve.
2.The aortic valve is separated from the tricuspid valve and right ventricular inflow by a prominent intervening tissue which comprises a subaortic conus which leads to the absence of tricuspid to aortic valve continuity.There is no sush conal tissue below pulm.valve as a result there is direct continuity between the hinge points of AMVL and post cusp of pul.valve.

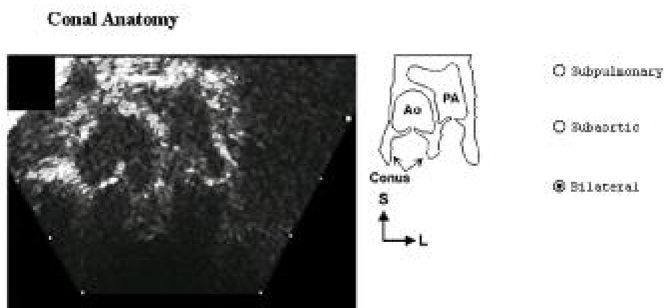






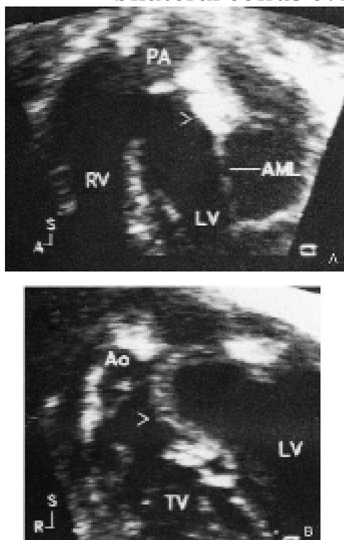
Subcostal view of an infant with d-transposition of the great arteries and a subpulmonic conus only. A: Subcostal coronal view demonstrates the posterior aorta (Ao), anterior and superior pulmonary artery (PA), and a subpulmonic infundibulum (Inf). There is ventricular arterial discordance. B: Subcostal coronal view swept further anteriorly demonstrating the anterior relationship of the pulmonary artery to the aorta. C: Subcostal oblique view showing a small subaortic ventricular septal defect (arrowhead). I, inferior;LPA, left pulmonary artery; lV, left ventricle; P/S, posterior/superior; RA, right atrium; RPA, right pulmonary artery; RV, right ventricle.

- *Bilateral*:Bilateral persistence of the subarterial conus usually results in double-outlet right ventricle.



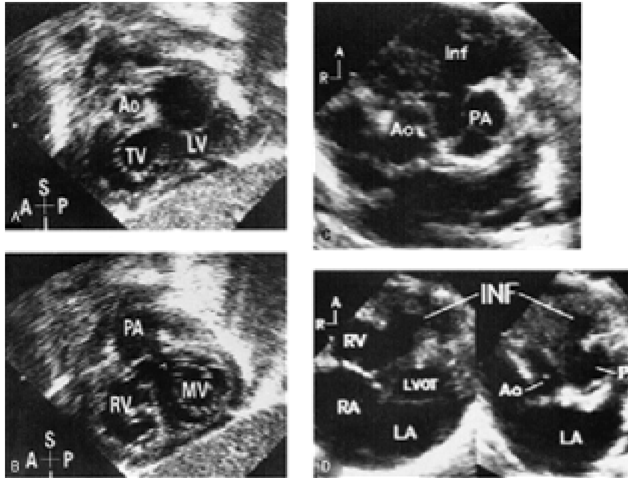
Bilateral conus:
Subcostal long axis sweep(in patient with double outlet right ventricle)
1.presence of conal tissue beneath both similunar valves as a result there is no continuity between AV valves and either similunar valve.This echo seen with bilateral conus.

- Particularly important is to determine which conus has the interventricular septum as one of its walls, to localize the ventricular septal defect with respect to the conus, and to determine the connection of the conus to the aorta.
- When two conuses are present, their relationship may be classified as either anterior/posterior or side-by-side *With the anterior/posterior conal relationship, the ventricular septal defect is usually subaortic; with the side-by-side relationship, the defect is usually subpulmonic*. The conal relationship can be determined by subcostal coronal and sagittal imaging with anterior/posterior and left/right sweeping, respectively. With an anterior/posterior conal relationship, the outlet septum inserts anteriorly separating the anterior conus from most of the interventricular septum and, thus, the ventricular septal defect. With a side-by-side conal relationship, the outlet septum inserts near the crux of the heart separating the lateral conus from the interventricular septum and ventricular septal defect.
- Bilateral conus also can be associated with d-transposition of the great arteries with VSD, a more anterior and superior location of the pulmonary root than in patients with a subaortic conus only, and discontinuity between the pulmonary and mitral valves (as well as between the aortic and tricuspid valves) .
- L-transposition of the great arteries also can be associated, although rarely, with bilateral conus evident in the subcostal views.



Subcostal sagittal views in a patient with transposition of the great arteries with a bilateral conal relationship.
A: Conal tissue (arrow) separates the pulmonary valve from the anterior leaflet of the mitral valve (AML).
B: Conal tissue (arrow) separates the aortic valve (Ao) from the tricuspid valve (TV). A,anterior; LV, left ventricle; PA, pulmonary artery; RV, right ventricle; S, superior.

- *Absent:*A rare type of d-transposition can exist in the context of bilaterally deficient subarterial conus. This results in an unusual heart in which d-transposition of the great arteries exists with a *doubly committed VSD and a posterior aorta*. These relationships can be identified by parasternal and subcostal coronal and sagittal imaging .

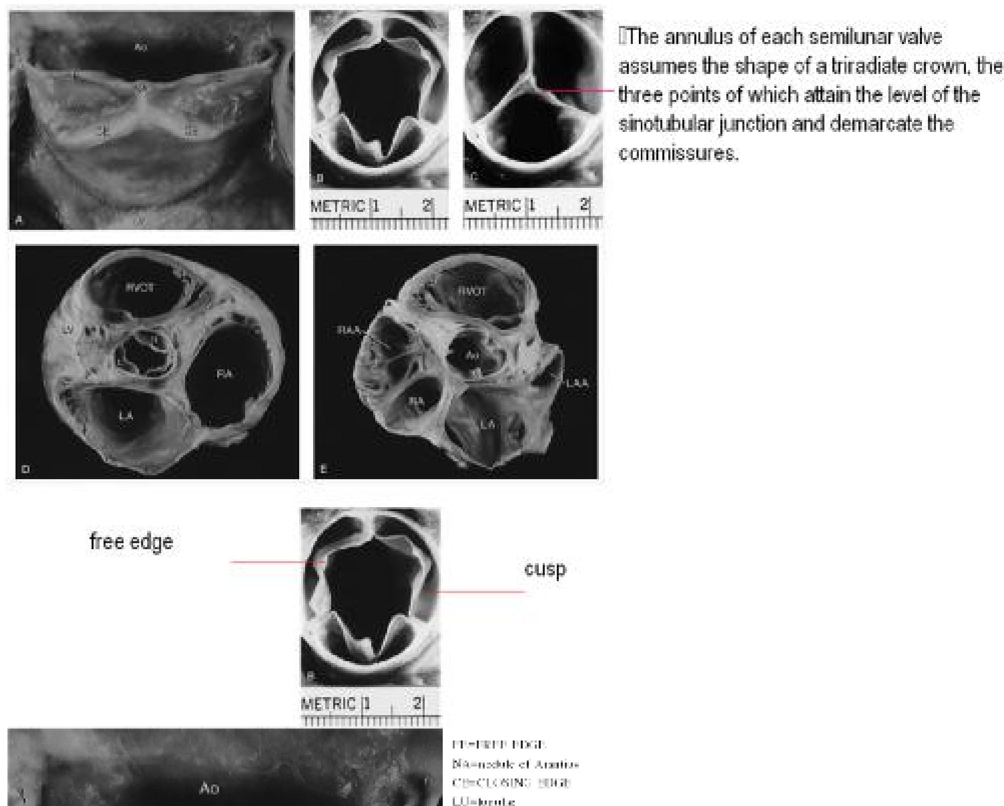


An infant with transposition of the great arteries, doubly committed ventricular septal defect, posterior aorta, and bilaterally deficient subarterial conus. A: Subcostal sagittal view demonstrating continuity between the aortic valve (Ao) and the tricuspid valve (TV). B: Subcostal sagittal view swept slightly leftward from A demonstrates continuity (arrow) between the pulmonary valve and the mitral valve (MV). C: Parasternal short-axis view showing the great vessel relationship with the aorta slightly posterior to the pulmonary artery (PA). There is an infundibular chamber (Inf) anterior to both semilunar valves. D:Parasternal short-axis view demonstrating the relationship of the infundibular chamber to the right ventricle and the left ventricular outflow tract (LVOT) immediately inferior to the pulmonary artery. A, anterior; I,inferior; LA, left atrium; P, posterior; S, superior.

Semilunar Valves

General Features

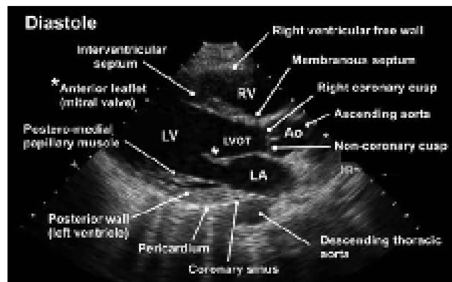
- The semilunar valves connect the ventricles to the great arteries and serve to maintain unidirectional blood flow.
- They consist of an annulus, cusps, and commissures. Because they have no tensor apparatus (tendinous cords and papillary muscles), the semilunar valves are simpler than the atrioventricular valves, and their opening and closure are primarily passive processes.
- Behind each cusp is an outpouching of the great artery that imparts a tribulbous, or cloverleaf, shape to the arterial root. The junction between the sinus portion of a great artery and its distal tubular portion forms a prominent ridge, the *sinotubular junction*. From the right and left aortic sinuses, proximal to this junction, arise the right and left coronary arteries, respectively. These are often incorrectly called the right and left coronary sinuses; the coronary sinus, of course, is a venous structure that empties into the right atrium.
- *The annulus of each semilunar valve assumes the shape of a triradiate crown, the three points of which attain the level of the sinotubular junction and demarcate the commissures.*
- A commissure, in turn, represents the site at which two cusps meet along the annulus(so is seen in diastole).
- As half-moon shaped (semilunar) structures, the cusps represent pocketlike flaps of delicate fibrous tissue. The leading edge of each cusp is its free edge, beneath which lies a shallow biscalloped ridge, the closing edge, along the ventricular surface of the cusp (Fig. below). At the center of each cusp, along the free edge, is a small fibrous mound, the *nodule of Arantius*. To either side of this nodule, between the free and closing edges, are two crescent-shaped areas called lunulae that represent the contact surfaces between adjacent cusps during valve closure. The arterial surface of each cusp, in conjunction with its arterial sinus, forms the valve pocket.



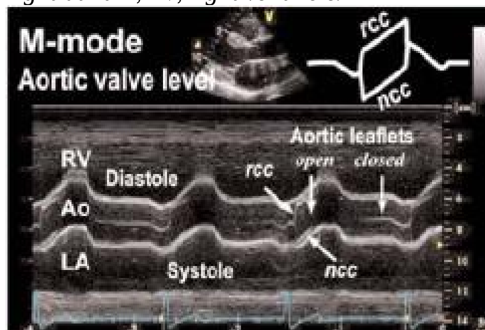
Semilunar valves. A: Each cusp is pocket shaped and has a free edge, a closing edge, a nodule of Arantius, and two contact regions (lunulae). The annulus (dotted line) for each cusp is U shaped. B and C: The aortic valve, viewed from above, is shown in simulated opened (B) and closed (C) positions. D and E: Tomographic sections at the level of the aortic valve show adjacent structures as viewed from above (D) and below (E).
Ce=closing edge, NA, nodule of Arantius, FE, free edge



Short axis through the aortic valve, visualized in the center of the screen, is comprised of three cusps, the right coronary cusp (RCC), the non-coronary cusp (NCC) and the left coronary cusp (LCC). Anterior to the aortic valve is the right ventricular outflow tract, with the tricuspid valve seen at 10 o'clock, and the pulmonic valve seen at approx 2 o'clock to 3 o'clock. The left atrium is immediately posterior to the aortic valve in this view.



Annotated parasternal long-axis view (PLAX) (depth 15–16 cm). Ao, aortic root; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.



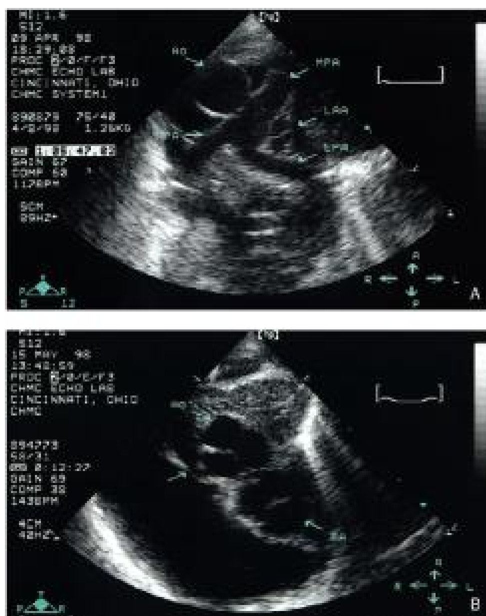
Aortic valve(M-mode)

Great Arteries

- The great arteries are formed by complete separation of the developing conotruncus; by the aortopulmonary septum.
- Normally, the aortic valve is posterior and rightwards of the pulmonary valve.
- There is fibrous continuity between the aortic and mitral valves due to absence of subaortic conus .
- Conversely, the pulmonary valve is lifted anteriorly, superiorly and leftwards by the subpulmonary infundibulum. There is no continuity between the pulmonary valve and the A-V valves.
- The great arteries intertwine as they leave the heart. Thus, the main pulmonary artry warps around the ascending aorta as it heads posteriorly, and the right pulmonary artery runs rightwards behind the ascending aorta.

Relationship:

- The aorta courses superiorly toward the thoracic inlet before coursing posteriorly, gives rise to strap vessels as it courses posteriorly, and has coronary arteries arising from its root.
- The pulmonary artery courses posteriorly almost immediately after it arises from the heart and bifurcates shortly after its origin. *The most helpful views for identifying the great vessels are the parasternal short-axis view at the base and even more superior, the suprasternal notch long- and short-axis views and the subcostal coronal and sagittal views .*



Parasternal short-axis images demonstrating different relationships between the great vessels. A: An echocardiogram from an infant with tetralogy of Fallot and pulmonary atresia demonstrates the identifying features of a pulmonary artery (MPA) with its bifurcation into right and left pulmonary arteries (RPA, LPA). The great vessels are normally related. B: In a patient with d-transposition of the great arteries,both semilunar valves are in cross section in the parasternal short-axis view. The aorta is identified as the rightward anterior vessel and has a right coronary artery (arrow) originating from the right facing sinus of Valsalva. A, anterior; Ao, aorta; LAA, left atrial appendage; P, posterior; PA, pulmonary artery.

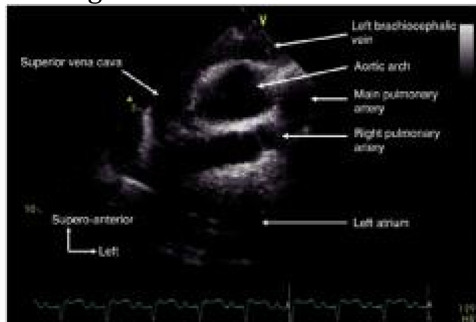
- *Situs solitus* of the great vessels describes the normal relative position of the *aortic annulus* located *rightward and posterior* to the pulmonary annulus. This relationship is best seen in the parasternal short-axis view, but it is also obvious in the parasternal long-axis sweeps and the subcostal coronal and sagittal views.
- *Situs inversus* of the great vessels describes the relative position of the *aortic annulus* located *leftward and posterior* to the pulmonary valve annulus in dextrocardia. In this relationship, there is no transposition (i.e., the aorta continues to arise from the left ventricle and the pulmonary artery from the right ventricle). This relationship is evident in a right parasternal short-axis view and in the subcostal coronal and sagittal sweeps.
- *Transposition of the great arteries* is diagnosed when there is a discordant ventriculoarterial relationship. (By definition, then, it is impossible to diagnose double-outlet right ventricle with transposition.) Instead, this is referred to as *double-outlet right ventricle with malposition of the great vessels*.
  - *Transposition of the great arteries can exist with the aorta right and anterior (d), left and anterior (l), and directly anterior (A) to the pulmonary artery. In addition, the aorta may exist side by side or even posterior to the pulmonary artery. These relationships are best diagnosed in the basal short-axis and subcostal coronal and sagittal views.*

Echo Tips:

- Best shown from subcostal long and parasternal short at sweeps.
- The spatial relationship of the aortic valve to the pulmonary valve must be defied.
- Normal conal anatomy (presence of subpulmonary conus, absence of subaortic cons) must be demonstrated echocardiographically.

Main and Branch Pulmonary Arteries

- The main and branch pulmonary arteries are best seen in the basal (and more superior) short-axis and subcostal coronal and sagittal views.
- In addition, the right pulmonary artery is best seen in the suprasternal short-axis view. The left pulmonary artery usually can be seen when sweeping the standard suprasternal long-axis view to the left.
- Pulmonary valve stenosis is evident in the parasternal basal short-axis view. Frequently, continued clockwise rotation of the transducer can yield an en face view of the pulmonary valve.
- Supravalvar pulmonary stenosis is best visualized in this view.
- Aortopulmonary window is usually evident on the parasternal short-axis view and careful, deliberate sweeps between the aorta and main pulmonary artery in the subcostal coronal and sagittal views.



Suprasternal notch short-axis view demonstrating typical normal vascular anatomy. From this view the left brachiocephalic vein, superior vena cava, transverse aortic arch, main pulmonary artery, right pulmonary artery, and left atrium can all be visualized.

Aorta and Branch Vessels

- The aorta, which lies more posterior in the center of the heart, can be visualized in many different views,including the parasternal long- and short-axis views, the apical five-chamber view, the subcostal views, and the suprasternal notch views .
- The aortic arch is best seen in the subcostal oblique view and the suprasternal views.
- Valve morphology in aortic stenosis can be understood by examining the parasternal views. The short-axis views allow determination of dysplasia and number of leaflets.
- Supravalvar stenosis and root dilation are best measured in the parasternal long-axis view.
- The side of the aortic arch (important in tetralogy of Fallot, truncus arteriosus, hypoplastic left heart, vascular rings, and before tracheoesophageal fistula repair) is diagnosed by sweeping the transducer in the suprasternal long-axis view and noting the relationship of the arch to the trachea, the rings of which resemble a stack of coins. Equally important is sweeping the transducer in the suprasternal notch short-axis view from the origin of the aorta superiorly toward the arch and branch vessels and then back inferiorly and posteriorly following the descending aorta. Using this view, the transducer should also be swept superiorly to follow the course of each branch vessel arising from the arch.
- *In a normal*, *left-sided aortic arch*, the first branch vessel is a right brachiocephalic artery that can be shown to bifurcate into right subclavian and carotid arteries (Fig. below).

Suprasternal notch short-axis view swept superiorly to demonstrate the bifurcation of the brachiocephalic artery into right subclavian artery and right common carotid artery. The fact that the first strap vessel is a brachiocephalic artery that bifurcates into the right subclavian and common carotid arteries indicates that this patient has a left aortic arch.

- In a right-sided aortic arch, the first branch vessel is usually a left brachiocephalic artery, which usually bifurcates into a left subclavian and carotid arteries. In either case, the first branch vessel should be followed to its bifurcation. If no bifurcation is present, anomalous origin of the ipsilateral subclavian artery should be suspected.
- Likewise, superior sweeping from the origin of each of the other strap vessels upstream should be performed deliberately to completely delineate the strap vessel anatomy.
- *Arch hypoplasia, coarctation, and interruption* are best diagnosed from the suprasternal long- and short-axis views. The proximal (the segment between brachiocephalic and common carotid artery origins) and distal (the segment between common carotid artery and subclavian artery origins) transverse arch should be imaged and measured.
- *Aortopulmonary collateral vessels* (important in tetralogy of Fallot and single ventricle/Fontan physiology) are best seen in the suprasternal notch long- and short-axis views. The subcostal oblique view of the descending aorta also allows identification of these vessels.

Coronary Arteries

- The coronary arteries are best seen in the parasternal short-axis views.
- The coronary arteries can also be seen in the apical four-chamber view as they course in the atrioventricular grooves.
- The midportion of the right coronary artery is seen in the subcostal coronal view.
- Frequently, the origin of the left coronary artery and the left anterior descending coronary artery are seen in the subcostal coronal view or a leftward sweep from the standard parasternal long-axis view.
- Anomalous origins and courses are important to delineate in
    - transposition of the great arteries, tetralogy of Fallot,
    - anomalous origin of the left (or more rarely, the right) coronary artery from the pulmonary artery,
    - dilated cardiomyopathy,
    - and exertional syncope

and are seen in the parasternal short axis.

- If 2-D imaging in the short-axis view shows *isolated dilation* of a proximal coronary artery, a coronary artery fistula or anomalous origin of a coronary artery should be suspected.
- The entire extent of the coronary artery must be interrogated for fistulous communications from all imaging planes.

- The main and branch pulmonary arteries should be carefully imaged for an anomalously arising coronary artery. *Simultaneous use of color Doppler is extremely helpful when investigating for coronary artery fistulae or anomalous origin of a coronary artery*.
- Coronary cameral sinusoids, seen within high-pressure ventricles such as the right ventricle in pulmonary atresia with intact ventricular septum, are best visualized in the apical four-chamber view with simultaneous use of color Doppler.
- Evaluation of the coronary arteries during and following Kawasaki's disease for aneurysms and stenoses should be performed in all the aforementioned imaging planes so that almost the entire extent of the coronary artery is interrogated.

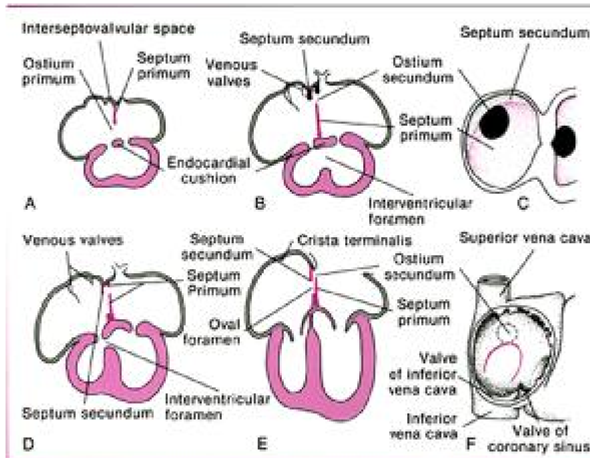
Chapter 4

The Interatrial Septum

The Interatrial Septum

- Defects of the atrial septum area common form of congenital heart disease.
- They may occur as isolated defects, or in association with other defects.
- The nomenclature of these defects is based on defect location within the atrial septum and the embryologic basis (origin.) Secundum ASD is the commonest form of atrial septal defect.

Patent Foramen Ovale
Secundum ASD
Primum ASD
Common Atrium
Sinus Venosus - SVC type
Sinus Venosus - IVC type



Diagrammatic representation of the atrial septa at 30 days (A), at 33 days (B), at 33 days (seen from the right side) (C), at 37 days (D), and in the newborn (Ey, the newborn atrial septum viewed from the right (F). (From Clark, E.B., and Van Mierop, L. H. S.: Development of the cardiovascular system. In Moss'Heart Disease in Infants, Children, and Adolescents. Baltimore, © Williams and Wilkins, 1989.)

Patent Foramen Ovale

- The foramen ovale is normally patent during fetal life and is bordered by the limbus of the fossa ovalis.
- It is covered by the flap valve of suptum primum and is patent in the fetus. After the fetus is born, left atrial pressure exceeds right atrial pressure, and the foramen ovale usually undergoes functional closure.
- If the foramen ovale is not anatomically closed, the potential for right.-to-left. shunting exists whenever right atrial pressure exceeds left atrial pressure.
- Atrial dilation can also lead to the potential for interatrial shunting across the foramen ovale.
- 20% of adults may have intermittently patent fossa ovale (PFO).

Echo Tips:

- Best shown from subcostal long and short axis views.

- May be very small, and in large subjects, may be shown only with color flow Doppler.

Should be measured in two orthogonal planes at end-diastole using 2-D imaging as well as color.

- Direction of flow (left-ro-right, right-to-left or bidirectional) should he assessed using color flow and pulsed wave Doppler.
- Redundant (aneurysmal) interatrial fossa tissue carries a higher likelihood of PFO.



Subcostal color doppler of the thin region of the fossa ovale showing the small left-to-right shunt at its edge.



Subcostal 2D view of the thin region of the fossa ovale.

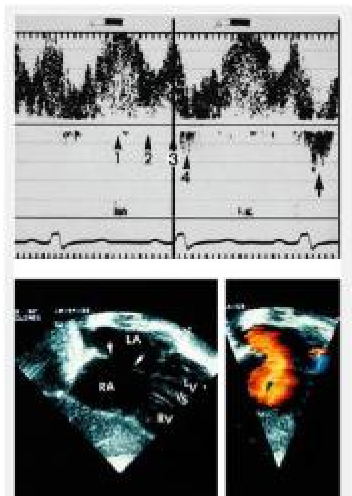
Secundum Atrial Septal Defect

These defects are due to :

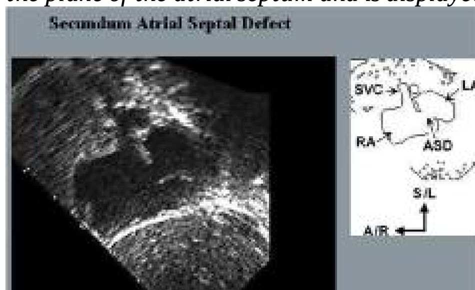
- deficiency of septum primum (the flap valve of the fossa ovalis),
- deficiency of septum secundum
- or both.
- Persistent strands of septum primium result in a fenestrated defect with left-to-right shunting across multiple orifices.
- Atrial-level left-to-right shunting across these defects can lead to right heart dilation.

Echo Tips:

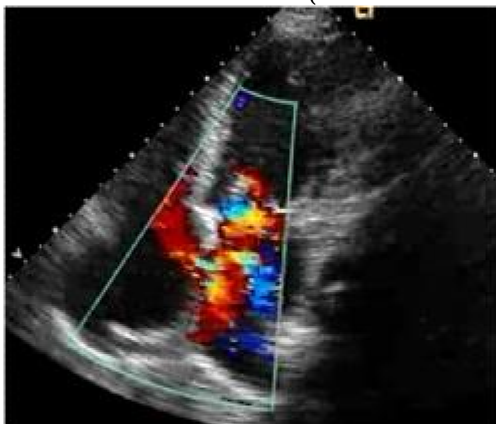
- Best shown from subcostal long and short axis views.
- Should be measured in two orthogonal planes at end- diastole using 2-D imaging as well as color flow (measurement of jet width).
- Defect location, margin and presence of malalignment of any component from atrial septum should be noted.
- Direction of flow (left-to-right, right-to-left or bidirectional) should he assessed using color flow and pulsed wave Doppler.



Pulsed Doppler echocardiographic findings in an atrial septal defect (ASD). The pulsed Doppler echocardiographic signal is consistent with a left-to-right shunt at the atrial level (recorded from a subcostal transducer position). The Doppler signal has a characteristic phasic change. Top: 1, left-to-right shunt (positive Doppler signal) begins in late systole; 2, diminishes through middiastole; 3, is enhanced by atrial contraction; and 4, reverses in early systole, consistent with a transient right-to-left shunt.
The early systolic right-to-left shunt is enhanced with inspiration (arrow) and diminishes with expiration. Bottom: Secundum ASD. Photograph of a color-flow Doppler echocardiographic image of a left-to-right shunt through a secundum ASD. Left:Subcostal four-chamber two-dimensional echocardiographic examination shows a large secundum ASD (arrows). Right: Left-to-right shunt at the atrial level is characterized as a velocity volume moving across the plane of the atrial septum and is displayed as an orange-red signal moving toward the transducer (arrow).



Moderate size secundum ASD(SUBCOSTAL VIEW)



Apical 4-chamber view shows the shunt from left atrium to right atrium.



Pulsed-wave doppler sampling at the right side of the 2nd ASD defect show a continuous left to right atrium flow with velocity of up to 1 m/s.

Primum Atrial Septal Defect

- Primum defects are seen anterior and inferior to the fossa ovalis.
- They are bordered by a cresent of atrial tissue superiorly (concavity facing inferiorly), and by the atrioventricular valves inferiorly. These defects may be larger in the anterior posterior dimension than in the superior-inferior extent.
- The septal attachment of the anterior mitral leaflet is apically displaced, leading the loss of the normal mitral-tricuspid valve offset. A cleft in the anterior mitral leaflet is frequently associated with this defect.

Echo Tips

- Best show n from apical and subcostal windows.

These defects should be measured in two orthogonal planes at end-diastole using 2-D imaging as well as color.

- Direction of flow (left-to-right, right-to-left or bi-directional) should be assessed using color flow and pulsed wave Doppler.
- The presence and degree of right sided volume overload must be assessed.
- Right venitricular pressure must be assessed, since pulmonary hypertension may occur with this defect.
- The location of this defect is close to the coronary sinus ostium. A large coronary sinus ostium (as may occur with persistent left superior vena cava to coronary sinus) may be mistaken for a primiun ASD unless multiple planes of inetrrogation are used.
- *Dense chordal attachments to the septal crest lead to the potential for subaortic stenosis, which may develop with time and should be sought by serial study.*
- The septal attachment of the anterior mitral leaflet is apically displaced, leading to loss of the normal A-V valve offset at the cardiac crux.
- A *cleft in the anterior mitral leaflet* is commonly associated and must be sought with a careful *parasternal short axis sweep*.

Common Atrium

- This anomaly is characterized by absence of most of the atrial septum.
- In the presence of two ventricles, common atrium is *always* associated with atrioventricular canal defect.
- This anomaly is usually due to coexistence of a large primum and a large secundum atrial septal defect.

- In addition, particularly in patients with visceral heterotaxy, there may be absence of other components; of the atrial septum, including the Coronary sinus and sinus venosus.

Echo Tips:

- Best shown in subcostal long axis and apical four-chamber views.

The sites of entry of the systemic and pulmonary veins into the common atrium must be identified.

Atrial Septal defect - Postop

- Repair consists of ASD closure utilizing direct suture approxirmation of the edges of the defect or pad closure using pericardium or prosthetic material.
- Post-op echocardiograms should assess for residual atrial-level shunts and right ventricular systolic pressure.
- Device closure should be assessed for site and leak.

Sino-septal defects

- Sino-septal defects refer to those that involve the area of the atrial septum derived from the sinus venosus.
- Most commonly this includes defects that occur at the junction of the superior vena cava and the right atrium, although these defects may also occur at the junction of the inferior vena cava and the right atrium.

Sinus Venosus ASD SVC Type

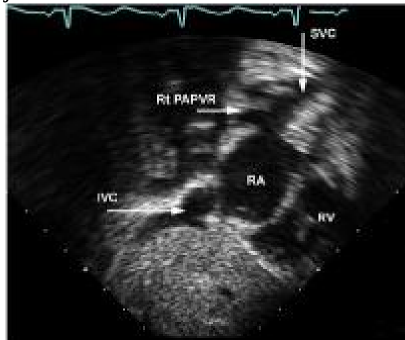
- These defects are located posterior and superior to the fossa ovalis.
- Normally, the right upper pulmonary vein enters the left atrium immediately adjacent to the Posterior superior aspect of the atrial septum .
- The anterior wall of the right upper pulmonary vein is normally shared with the *adjacent atrial septum*, *posterior wall of the superior vena cava and right atrial wall*.
- In this anomaly, the anterior wall of the right upper pulmonary vein is absent, resulting in anomalous drainage of the right upper pulmonary vein into the right atrium .
- If the defect is large, the right upper pulmonary vein drains into the lateral wall of the right atrium adjacent to the entrance of the superior vena cava.
- The right middle lobe may likewise have anomalous pulmonary venous drainage.

Echo Tips:

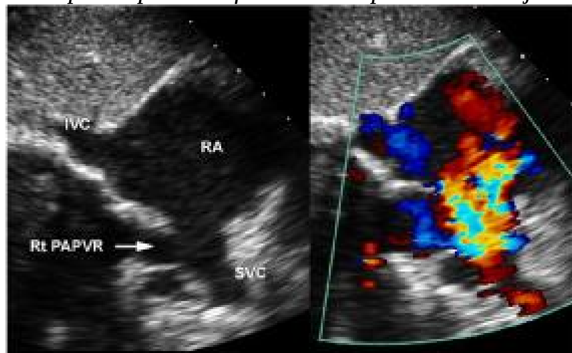
- Best shown in subcostal short axis view with extreme rightward angulation, tracking the course of the right upper pulmonary vein proximally towards the atria.
- The high right parasternal view in the sagittal plane is very useful in demonstrating the defect and its relationship to the superior vena cava.
- Pulmonary venous anatomy must be established: anomalous return of the right upper pulmonary vein to the junction of the right atrium and the superior vena cava is the rule.
- Should be measured in two orthogonal planes at end-diastole using 2-D imaging as well as color.
- Direction of flow (left-to-right, rigt-to-left or bidirectional) should be assessed using color flow and pulsed wave Doppler.



This shows the difficulty of visualizing a sinus venosus defect. The usual subcostal view (left image) seems to show an uninterrupted atrial septum. By tilting superiorly (right image) the sinus venosus becomes evident (arrows) near the junction with the SVC and an anomalous pulmonary vein which often accompanies these type of defects.



This (inverted) subcostal view is labelled to show partial anomalous pulm. veins (PAPVR) entering a sinus venosus defect in the superior portion of the atrial septum near the junction with the SVC.



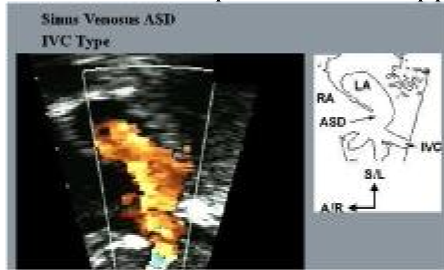
These subcostal images show relevant structures labelled (left image). The sinus venosus defect lies at the junction of the SVC-RA with a partial anomalous right pulmonary venous return (Rt PAPVR). Bloodflow patterns are shown in the color doppler image on the right.

Sinus Venosus ASD IVC Type

- These defects are located posterior and inferior to the fassa ovails.
- Normally, the right lower pulmonary vein enters the left atrium immediatlely adjacent to the posterior inferior aspect of the atrial septum. *The anterior wall of the right lower pulmonary vein a normally shared with the adjacent atrial septum and right atrial wall*. In this anomaly, the anterior wall of the right lower pulmonary vein is absent, resulting in anomalous drainage of the right lower pulmonary vein into tie right atrium. If the defect is large, the right lower pulmonary vein drains into the lateral wall of the right atrium adjacent to the entrance of the inferior vena cava. Likewise, the right middle lobe may have anomalous pulmonary venous drainge.

Echo Tips:

- Best shown in the subcostal transverse plane, sweeping from the inferior vena cava to the heart; this defect is posteriorly and inferiorly located, and my be missed if the sweep does not start in the transverse plane.
- Pulmonary venous anatomy must be establisbed; anomalous return of the right lower pulmonary vein to the junction of the right atrium and the inferior vena cava is the rule.
- Should be measured in two orthogonal planes at end-diastole using 2-D imaging as well as color.
- Direction of flow (left-to-right, riglnt-to-left or bidirectional) should be assessed using color flow and pulsed wave Doppler.



SUCOSTAL SAX:SUCOSTAL SAX with steep rightward angulation shows a large atrial septal defect with left to right flow .T he defect is located inferiorly and posteiorly in the atrial septum immediately posterior and superior to the enterence of inferior vena cava .The right lower pulmonoary vein drain into the right atrium.

Chapter 5

The Interventricular Septum

The Interventricular Septum

- VSDs are the commonest form of structural congenital heart disease.
- VSDs may occur in isolation or in association with other structural heart defects.
- VSDs vary in size, location and natural history.
  - Small VSDs in the membranous or muscular septum may close spontaneously.
  - Large defects may lead to left sided chamber enlargement or pulmonary hypertension, necessitating surgical closure.
- An echocardiogram in a patient with a VSD should assess the hemodynamic significance of the defect and the feasibility of surgical closure.

Membranous
Muscular
Canal-type (Inlet)
Supracristal (Conal-Septal)
Anterior Malalignment
Posterior Malalignment

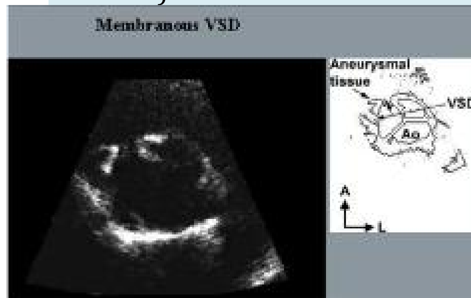
Membranous VSD

- (also known as 'conoventricular')
- These comprise the commonest form of VSD.
- When viewed from the left ventricle, these defects are located in the left ventricular outflow tract inferior to the aortic valve. When viewed from the right ventricular aspect, these defects lie beneath the crista supraventricularis and posterior to the papillary muscle of the conus.
- *The tricuspid valve may be intimately involved in the defect: so-called aneurysmal tissue formation, consisting of fibrous tag of tricuspid valve tissue, can lead to partial or complete closure of the defect.*

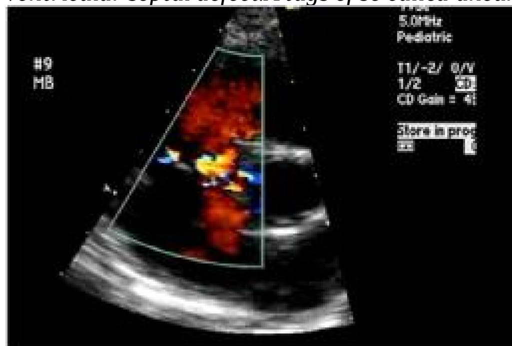
Echo Tips:

- Best shown in parastertal short axis (between 9 and 12 o'clock), apical and subcostal views.
- Tricuspid valve tissue (aneurysmal tissue) may lead to varying amounts of flow: the color flow jet across the VSD should be measured.
- Defect location, margins and event should be determined.
- Sizing of the defect should be performed in the widest dimension at end-diastole in orthogonal planes using 2-D imaging as well as color.
- Chordae or papillary muscles straddling the defect should be identified.
- Structures that may impede surgical visualization of the entire extent of the defect, such as prominent right ventricular muscle bundles, should be noted.
- The potential for outflow tract obstruction due to malalignment of the infundibular septum must be recognized.

- If there is evidence of elevated right ventricular presssure, the color sweep of the ventricular septum that is routinely performed to rule out VSDs should utilize a low color scale (nyquist limit).



PSX view is importent to differentiate among types of VSDs
This PARASTERNAL SAX shows an echo free space in the region normally occupied by the membranous septum ,immediately anterior and inferior to the septal leaflet of the tricuspid valve.This represent a small membranous ventricular septal defect.A tags of so called aneurysmal tissue from the tricuspid valve is seen partially occluding the VSD.



Color Doppler SAX



Four chamber view shows the membranous VSD with aneurysm formation

Muscular VSD

Defects may occur in the muscular portion of the trabecular ventricular septum, inferior to the septal band or superior to the septal band. These are referred to as muscular ventricular septal defects. They may be singular and small or alternatively can be multiple with significant hemodynamic effects. The former tend to have a high incidence of spontaneous closure, while the latter can be complicated, an in the most severe manifestation, give a "Swiss cheese" appearance to the ventricular septum.

- These defects are frequently multiple.

- They are usually best seen from the left ventricular aspect,where the ventricular septum is smooth; overlying muscular trabeculations usualIy make these defects difficult to visualize from the right ventricular aspect,
- These trabeculations way divide a VSD into two or more orifices on the right ventricular aspect. The occurence of multiple closely-spaced defects can lead to a *so-called "Swiss cheese" appearance* of the ventricular septum posing a techical challenge to the surgical closure of these defects.
- *Spontaneous closure of small and moderate sized muscular VSD's frequently occurs in the first two years of life*.

Echo Tips:

- Multiple muscular VSDs are common: careful base-to-apex sweeps using two-dimensional imaging and color flow Doppler must be preformed to rule out additional VSDs.
- These VSDs may be serpiginous with more than one opening on the right ventricular aspect; each VSD should be visualised on the right as well as left ventricular aspects.
- Defect location, margins and extent should be determined.
- *Sizing of the defect should be performed in the widest dimension at end-diastole in orthogonal planes using 2-D imaging as well as color.*
- Chordae or papillary muscles straddling the defect should be identified. Structures that may impede surgical visualization of the entire extent of the defect, such as prorminent right ventricular muscle bundles, should be noted.
- The potential for outflow tract obstruction due to malalignment of the infundibular septum must be recognized .
- If there is evidence of elevated right ventricular pressure, the color sweep of the ventricular septum that is routinely performed to rule out VSDs should utilize a low color scale (Nyquist limit).



Apical 4 chamber view with color doppler imaging shows evidence of the small muscular VSD shunting from LV to RV.



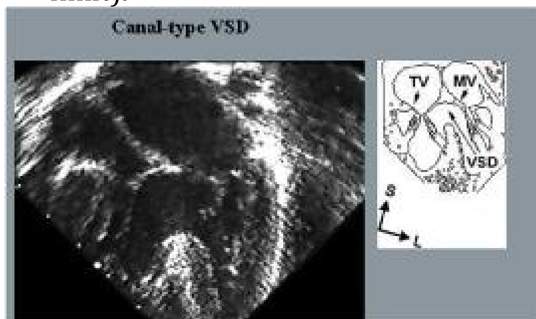
The CW doppler on the left shows the systolic pressure gradient from LV to RV is 90 mm Hg indicating that the VSD orifice is small and restrictive (implying a small volume shunt). The image on the right is the short axis LV view marked at the site of the small VSD.

Canal-type VSD(Inlet)

- These defects may occur in isolation or may be associated with defects of the atrial primum and be a component of the atrioventricular septal defect (to be discussed below). Patients with inlet ventricular septal defects may demonstrate a superior axis on electrocardiogram.
- Are known as artioventricular canal-type or atrioventricular septal defects.
- *These defects are located posterior and inferior to membranous VSDs beneath the septal leaflet of the tricuspid valve and inferior to the papillary muscle of the conus.*
- These defects are frequently large and associated with pulmonary pertension.

Echo Tips:

- Best shown from apical and subcostal views.These defects are frequently large, and may extend into the muscular or membranous septum.
- Tricuspid valve chordae straddling the defect may preclude VSD closure; these should be sought in each case.
- Other endocardial cushion defects, such as cleft anterior mitral valve leaflet and primum ASD, should be ruled out.
- Defect location, margins and extent should be determined.
- Sizing of the defect should be performed in the widest dimension at end-diastole in orthogonal planes using 2-D imaging as well as color.
- Chordae or papillary muscles straddling the defect should be identified.
- Structures that may impede surgical visualization of the entire extent of the defect, such as prominent right ventricular muscle bundles, should be noted.
- The potential for outflow tract obstruction due to malalignment of the infundibiilar septum must be recognized.
- If there is evidence of elevated right ventricular pressure, the color sweep of the ventricular septum that is routinely performed to rule out VSDs should utilize a low color scale (nyquist limit).



The Interventricular Septum-Canal-type (Inlet)
This APICAL view angling posterior to the plane of the AV valves shows a large defect in the posterior superior ventricular septum .This defect extends to the cardiac crux and represent an AV canal type ventricular septal defect.

Supracristal (Conal - Septal)

- A small portion of VSDs occur in the outflow portion of the right ventricle above the crista supraventricularis.
- *These are known as doubly committed subarterial, conoseptal hypoplasia or supracristal VSDs.*
- These defects are directly beneath the aortic valve on the left ventricular side of the septum and open below the pulmonary valve in the infundibulum on the right ventricular side.

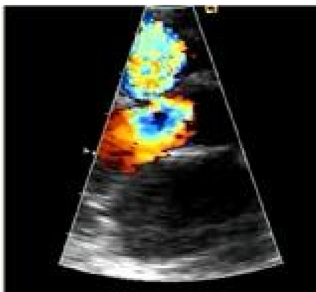
- The defect is many times associated with aortic valve prolapse, which may lead to aortic valve insufficiency.
- These defects are found more commonly in people of Asian decent.
- Ventricular septal defects, which are doubly committed subarterial and represent conoseptal hypoplasia, are usually well profiled from a parasternal long axis, short axis and subcostal view. *In the subcostal and short axis view, the defect will be seen in the right ventricular outflow tract.*
- It is important to thoroughly evaluate these patients for any evidence of aortic valve prolapse, specifically of the right coronary cusp, and for aortic regurgitation.

Echo Tips:

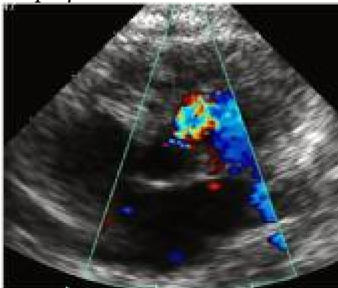
- Best shown from parasetnal short axis at (between 12 and 3o'clock} and parasternal long axis.
- *Frequently associated with prolapse of right coronary cusp of the aortic valve in to the defect which may partially close the defect and may also cause aortic valve insufficiency: aortic valve cusp mobility and coaptation should be assesed.*



Long axis of the left ventricular outflow tract shows the ventricular septal defect just below the right coronary cusp of the aortic valve.



Color doppler of the left ventricular outflow tract shows the ventricular septal defect just below the right coronary cusp of the aortic valve.



Color doppler of the left ventricular outflow tract in short axis shows the ventricular septal defect at 1 o'clock just below the pulmonary valve and adjacent to the right cusp of the aortic valve. This is characteristic of a supracristal VSD, distinguishing it from perimembranous VSDs.

Anterior Malalignment

- *This type of VSD is classically seen in Tetralogy of Fallot.*
- It is due to anterior leftward and superior deviation of the conal septum, which raises the 'floor' of the right ventricular outflow tract, causing subpulmonic stenosis.
- Since growth within the heart is related to blood flow patterns, it follows that subpulmonary obstruction can be associated with deficient growth of the pulmonary anulus and branch pulmonary arteries.

Echo Tips:

- Best shown from subcostal short axis view with anterior angulation and clockwise rotation of the transduser.
- Note Ê *In the presence of a VSD, Doppler gradient across the outflow tracts are an insensitive method for assessing the potential for post-operative outflow tract obstructon.*

Posterior Malalignment

- *This type of VSD is classically seen in association with interrupted aortic arch.*
- It is due to posterior, rightward and inferior deviation of the conal septum into the left ventricular outflow tract causing subaortic stenosis.
- A variable degree of conal septal hypoplasia is an associated finding.
- Since growth within the heart is related to blood flaw patterns, it follows that subaortic obstruction can be associated with deficient growth of the aortic anulus, ascending aorta and aortic arch.

Echo Tips:

- Best shown from parastanal bong axis, apical views and base-to-apex short axis.

Frequently associated with hypoplasia of the infundibular septum.

VSD - Postop

Repair consists of closure, preceded by PA banding in rare cases.

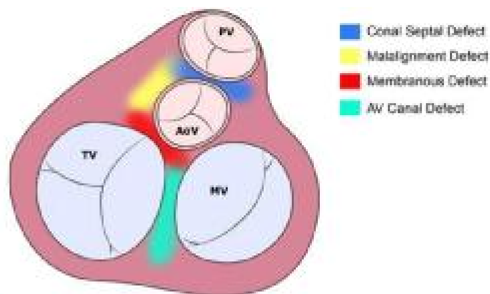
Echo foIIowing VSD closure:

- Residual VSD: All patch margins should be assessed, and the defect should be sized.
- Posterior inferior leaks are common following membranous VSD closure.
- Outflow tract obstructions: may he unmasked after VSD closure If tricuspid valve leaflets have to be detached for visualizing the VSD, valve regurgitation or stenosis may appear anew after surgery.
- Patch dehiscence: may be evident as free-floating VSD patch at one of its margins.

Echo following PA band;

- Band location in MPA, distance from pulmonary valve and from MPA bifurcation: encroachment on branch PAs.
- Peak gradient across the band.
- Band erosion into MPA.

Summary of echo findings:



Planes of Doppler interrogation for different ventricular septal defects as seen in parasternal short axis. PV: Pulmonary valve; AoV: Aortic valve; TV: Tricuspid valve; MV:Mitral valve.

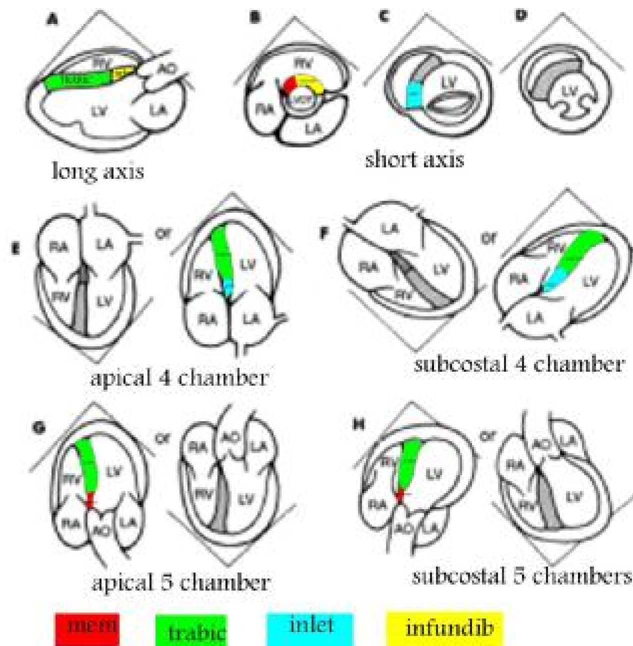


Illustration of various types of VSD

Chapter 6

Atrioventricular Canal Defects

Atrioventricular Canal Defects

- The endocardial cushions proliferate within the developing A-V canal and subdivide the common A-V valve orifice into tricuspid and mitral components; the cushions also participate in normal formation of both A-V valves.
- In addition, these cushions close ostium primum and the interventricular foramen.
- Defects of the A-V canal are due to partial or complete failure of fusion of the superior and interior endocardial cushions, leading to partial or complete A-V canal defects respectively.
- Atrioventricular septal defects (AVSD) result from abnormal development of the membranous and muscular atrioventricular septum. Depending on severity, the defect can be classified as complete or partial.
- The ventricular septal defect is located at the 'inlet' portion of the ventricle and the atrial septal defect occurs as an ostium primum type (a deficiency of the septum primum in its inferior and anterior aspect).
- AVSD can accompany other complex congenital heart disorders.

ASD Primum
Cleft Mitral Valve
Complete AV Canal,
Rastelli A Complete AV Canal,
Rastelli B Complete AV Canal,
Rastelli C Unbalanced CAVC

Primum Atrial Septal Defect

- These defects are seen anterior and inferior to the fossa ovalis, bordered by a crescent of atrial tissue superiorly (concavity facing inferiorly), and by the atrioventricular valves inferiorly.
- These defects may be larger in the anterior-posterior dimension than in the superior-inferior extent.
- The septal attachnment of the anterior mitral leaflet is apically displaced, leading to loss of the normal mitral-tricuspid valve offset. A cleft in the anterior mitral leaflet is frequently associated with this defect.

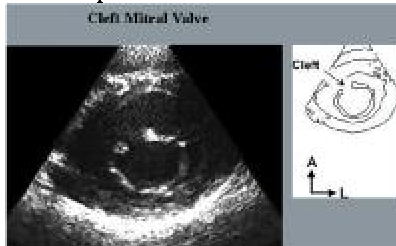


Subcostal view of the ASD primum . Cleft mitral valves are common in this entity.

CLEFT MITRAL VALVE

Echo Tips:

- Best shown from parasternal short axis view, sweeping from base to apex.
- Frequently associated with mitral regurgitation through the cleft; the presence and severity of MR are detected with color flow Doppler.
- The width of the regurgitant jet at its origin is best shown by color flow Doppler in the parasternal short axis sweep.



Cleft Mitral Valve:
This PARASTERNAL SAX sweep shows agap or a defect in the anterior mitral leaflet which represents a cleft.Note the typical location of the cleft in the middle of the leaflet.

Postop

- Surgery consists of suture closure of the cleft in order to prevent or treat mitral regurgitation.
- Post-operative echocardiograms: should assess for residual mitral regurgitation and also for mitral stenosis; the latter can occur if the underlying papillary muscle structure is abnormal.
- Thus, a single left ventricular papillary muscle can potentially result in a parachute mitral valve following closure of the cleft.
- Mitral stenosis can also occur if surgical closure of the cleft results in a small effective mitral valve orifice.

Complete Atrioventricular Canal

Rastelli type

- This defect is characterized by a
  - primum atrial septal defect,
  - a contiguous *inlet ventricular septal* defect and
  - a common atrioventricular valve.
- The atrial septum is crescentic with the concavity directed inferiorly, and the ventricular septum is crescentic with the concavity directed superiorly.
- The common atrioventricular valve usually has two bridging leaflets and three lateral leaflets. The underlying papillary muscle arrangement is variable; commonly, there are two papillary muscles in the left ventricle and three in the right ventricle.
- *Complete AV canal defects are classified (Rastslli classification) based on the morphology of the anterior bridging leaflet of the common AV valve and its attachments to the ventricular septum.*

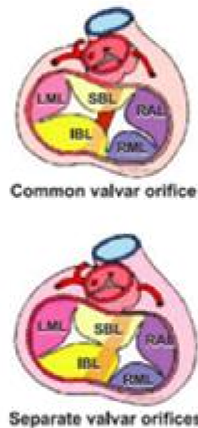
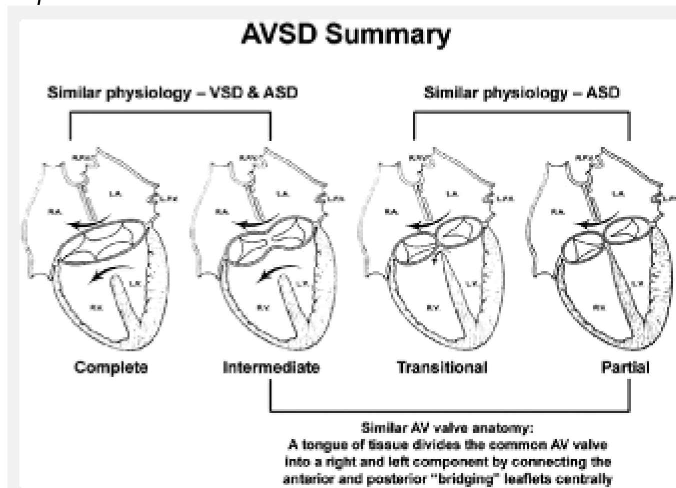


Diagram showing the common atrioventricular junction in hearts with atrioventricular septal defect. The junction is guarded by a valve with common orifice (upper panel) or separate right and left valvar orifices (lower panel). The common valve has five leaflets. Two of them, the superior and inferior bridging leaflets, are shared by both ventricles. Fusion between the bridging leaflets divides the common valvar orifice into two discrete orifices.IBL = inferior bridging leaflet, LML = left mural leaflet, RAL = right antero-superior leaflet, RML = right mural leaflet, SBL = superior bridging leaflet.



Summary of AVSD. Anatomic and physiologic similarities between the different forms of atrioventricular septal defect (AVSD) are illustrated. Complete AVSDs have one annulus with large interatrial and interventricular communications. Intermediate defects (one annulus, two orifices) are a subtype of complete AVSD. Complete AVSDs have physiology of ventricular septal defects (VSD) and atrial septal defects (ASD). In contrast, partial AVSDs have physiology of ASDs. Transitional defects are a form of partial AVSD in which a small inlet VSD is also present. Partial defects and the intermediate form of complete AVSD share a similar anatomic feature: A tongue of tissue divides the common atrioventricular valve into distinct right and left orifices. LA, left atrium; LPV, left pulmonary vein; LV, left ventricle; RA, right atrium; RPV, right pulmonary vein; RV, right ventricle

Rastelli type A

- Is characterized by a divided anterior leaflet that is attached to the crest and right side of the ventricular septum at the papillary muscle of the conus.
- This defect is characterized by a primum atrial septal defect, a contiguous inlet ventricular septal defect and a common atrioventricular valve. The atrial septum is crescentic with the concavity directed inferiorly, and the ventricular septum is crescentic with the concavity directed superiorly. The common atrioventricular valve usually has two bridging leaflets and

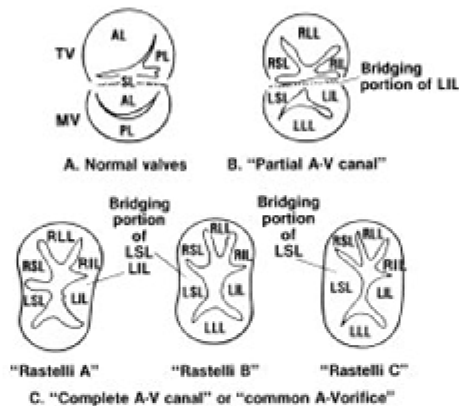
three lateral leaflets. The underlying Papillary muscle arrangement is variable; commonly, there are two papillary muscles in the left ventricle and three in the right. ventricle.

Rastelli type B

Rastelli type B: is characterized by a divided anterior leaflet that is attached to the right side of the ventricular septum

Rastelli type C

Rastelli type C: is characterized by an undivided anterior leaflet that is unattached to the ventrlcular septum; the right -sided attachments of the anterior leaflet are confined to the right ventricular free wall. This type of defect is common in patients with visceroatrial heterotaxy.



Echo Tips:

- Best shown from apical and subcostal views. *Chordal attachments of the common AV valve are best shown from sub costal short axis view with anterior angulation and clockwise rotation, the so-called `en-face' view .*
- Each component of the defect (prirmum ASD, common AV valve, canal-type VSD) should be defined.
- Sizing of atrial and ventricular septal defects should be performed in ***the widest dimension at end-diastole in orthogonal planes*** using 2-D imaging as well as color.
- Dense chordal attachments to the crest of the ventricular septum may decrease the effective shunt across a canal-type VSD; the width of the color Doppler jet across the VSD must be measured.
- The common AV valve is frequently dysplastic and regurgitant; the presence and severity of AV valve regurgitation and the origin of the regurgitant jets must be identified.
- Accessory AV valve attachments to the left ventricular outflow tract and chordal attachments to the crest of the ventricular septum may lead to outflow tract obstruction.
- Adequacy (number and spacing)of the left ventricular papillary muscles must be confirmed.

Postop

The goals of surgical repair are closure of the atrial and ventricular septal defects (using one or two patches), division of the common atrioventricular valve into two competent and non-stenotic atrioventricular valves, and repair of associated defects. Postoperative echocardiograms should assess for

- A-V valve regurgitation or stenosis

- ü Residual ASD or VSD
- ü Subaortic stenosis.

Unbalanced Complete Atrioventricular Canal

- § Unbalanced atrioventricular canal defects are characterized by hypoplasia of one of the two ventricles.
- § These defects are due to inadequate or excessive rightward migration of the atrioventricular canal.
- § The common atrioventricular valve has chordal attachments connecting exclusively or predominantly into one ventricle. This leads to preferential blood flow into that ventricle; the ventricle which lacks chordal attachments is frequently hypoplastic.

Echo Tips:

- § Best shown from apical and subcostal views.
- § The dominant ventricle is frequently dilated and hypertrophic.
- § While this finding can lead to the visual impression of hypoplasia of the contralateral ventricle, this diagnosis should be based on measurements of ventricular mass and volume (whenever possible) and not merely based on size disparity of the ventricles.

Unbalanced Complete AV Canal - Postop

If biventricular repair is not possible due to severe hypoplasia of one of the ventricles, patients may need staged repair for completion of the Fontan operation. For details on staged surgery (Fontan operation), go to Chapter 31.

Patients who have severe common A-V valve regurgitation may not be able to be staged for the Fontan operation, and may need cardiac transplantation. For details on the post-transplant heart, go to Chapter 30.

Echocardiography Summary:

- § The five leaflets of the common atrioventricular valve (superior/anterior,right and left bridging, right superior, right mural, inferior/posterior bridging, and left mural) are best seen in a *right anterior oblique subcostal view* (midway between the coronal and sagittal views).
- § In this view, the presence or absence of a tongue of tissue connecting the two bridging leaflets should be identified first to allow the establishment of a *complete (absent connecting tissue)* or *partial (present connecting tissue) atrioventricular septal defect. Then the degree of bridging of the superior leaflet and its attachments are identified, allowing for Rastelli's classification* .



The subcostal oblique view (a hybrid between coronal and sagittal views) of a common atrioventricular valve. The five leaflets of the common atrioventricular valve are shown. The commissure of the superior bridging leaflet attaches to the inferior portion of the outlet septum (Rastelli type A). I, nferior; S, superior.

- *Straddling And Crisscross Connections* are seen in the apical four-chamber and subcostal views. A *straddling atrioventricular valve* (valve attachments to contralateral ventricle)(involves the anomalous insertions of chordae tendineae (tendinous cords) or papillary muscles into the contralateral ventricle) must be distinguished from *mere overriding* (valve annulus partially displaced over the ventricle) in these views.
- Crisscross atrioventricular relationships necessitate deliberate and slow sweeping of the transducer anterior and posterior in the subcostal coronal or superior and inferior in the parasternal short-axis views with careful, simultaneous observation of each valve's upstream atrium and downstream ventricle.

Chapter 7

Tricuspid Valve and Right Atrium

Tricuspid Valve and Right Atrium

The right atrium and tricuspid valve develop from:

- † the sinus venosus (the cardiac ends of the venae cavae and coronary sinus),
- † septum primum and secundum,
- † the primitive trabeculated atrium and
- † endocardial cushions.
- † The subvalvular apparatus of the tricuspid valve develops from right ventricular myocardium.

- § The function of the right atrium and tricuspid valve is to provide unobstructed and non-regargitant flow from the systemic veins into the right ventricle.
- § This, in turn, requires normal development of all of the components of the right atrium.
- § Thus, the systemic veins must fuse with the primitive atrium, and the atrial septurn and A-V canal must develop normally.
- § Defective development of any component can lead to congenital anomalies involving the right atrium and tricuspid Valve and affecting right ventricular inflow.

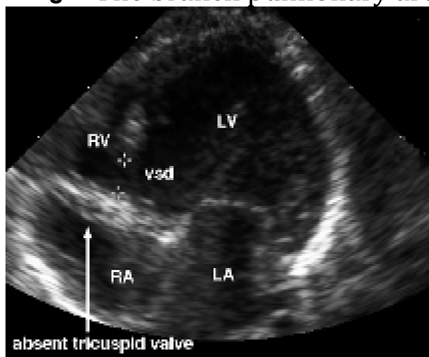
Tricuspid Atresia
Tricuspid Hypaplasia
Tricuspid Stenosis
Tricuspid Regurgitation
Ebstein Anomaly
Left Juxtaposition of Rt Atrial Appendage

Tricuspid Atresia

- § This is defined as agenesis of the tricuspid valve resulting in absence of any direct communication between the right atrium and right ventricle.
- § *Other cardiac anomalies are the rule*, includnig septal defects (an atrial septal defect is needed for survival), patent ductus arteriosus, d- or l-transposition of the great arteries and pulmonary or aortic outflow obstruction.
- § An atrial communication (ASD or PFO) must be present for systemic venous return to cross to the left atrium and be pumped from the left ventricle.
- § The net effect of these anomalies may lead to decreased, normal or increased pulmonary blood flow.
- § Relative or absolute hypoplasia of the right ventricle is an obvious consequence of the loss of direct AV connection. This can vary considerably, however, depending on the presence of a ventricular septal defect (VSD). A VSD will allow for entry of blood into the right ventricle and the pulmonary artery. If a VSD does not exist, the ventricle receives no flow and will be extremely small, with hypoplasia or atresia of the pulmonary valve and trunk.
- § If the ventricular septum is intact, a separate systemic arterial-to-pulmonary source is required such as a PDA or an aorto-pulmonary collateral vessel.

Echo Tips:

- Best shown from parasternal short axis and apical views.
- Characterization of segmental anatomy is essential; either ventricular d- or I-looping and similarly, either d- or l-malposition of the aortic valve may occur.
- All atrial and ventricular septal defects should be characterized and measured by echo.
- Spontanenus closure of ventricular septal defects frequently occurs, and therefore these defects should be measured serially.
- Left juxtaposition of the right atrial appendage is common in tricuspid atresia especially when associated with d-transposition of the great arteries.
- Aortic outflow obstruction can occur at multiple levels, most often seen in tricuspid atresia with d-loop ventricles and d-transposition of the great arteries.
- Pulmonary outflow obstruction can occur at multiple levels, most often seen in tricuspid atresia with d-loop ventricles and solitus arrangement of the great arteries (normal ventriculoarterial connections).
- The branch pulmonary arteries must be assessed.



Echo shows a '4 chamber view' of a newborn with TA, a VSD and a small RV.

Tricuspid Hypoplasia

- This is characterized by a small valve anulus and diminutive valve leaflets and chordae, with normal valve structure.
- This defect is frequently associated with right ventricular hypoplasia and pulmonary outflow obstruction especially if the ventricular septum is intact.

Echo Tips:

- Best shown from parasternal short axis and apical views.
- Valve anulus diameter should be measured in apical or parasternal short axis; plotting this on standard nomograms enables calculation of the z score.

Tricuspid Stenosis

- This may be congenital or acquired (rheumatic or secondary to infiltrating tumor).
- Valve anulus size is variable.
- The valve leaflets may be dysplastic and thickened; the chordae tendineae maybe fused or foreshortened, and the papillary muscles may be fused or closely spaced.

Echo Tips:

Since the venae cavae and the right atrium have high capacitance, dilation of these structures is seen before Doppler techniques can detect tricuspid stenosis.

Tricuspid Regurgitation

- This maybe due to either structural abnormalities of the tricuspid valve (Ebstein anomaly, elongated or ruptured chordae, or an unguarded tricuspid valve i.e., absence of valve tissue with a normal anulus), abnormal papillary muscle function (e g., perinatal asphyxia). or annular dilatation.

Ebstein Anomaly

- Normal mitral-tricuspid valve offset is less than 8 mm/m^2 body surface area.
- An increase in this offset indicates displacement of the septal leaflet of the tricuspid valve into the right ventricle, which becomes thin-walled and is continuous with the right atrium. This 'atrialization' of the right ventricle is characteristic of Ebstein anomaly.
- Tricuspid valve morphology is widely variable and may include variable degrees of tethering of the septal and posterior leaflets to underlying myocardium. The anterior leaflet may be redundant or fenestrated, and the valve anulus maybe dilated.
- Right ventricular myocardium may be thinned and dysfunctional, and ventricular, septal motion maybe paradoxical.
- Associated lesions, including atrial and ventricular septal defects, pulmonary valve stenosis or atresia, are common.

Left Juxtaposition of
Right Atrial Appendage

- This anomaly is frequently seen in association with complex heart disease such as d-transposition of the great arteries, particularly in association with tricuspid atresia.
- In this lesion, the tight atrial appendage is displaced leftwards while remaining in continuity with the right atrial cavity.
- The appendage passes behind the ascending aorta and comes to lie leftward of the ascending aorta, adjacent to and to the right of the left atrial appendage.
- The recognition of this anomaly is important for defining atrial septal anatomy accurately in these patients who frequently need procedures such as balloon atrial septostomy.
- As a result of the juxtaposition, the anterior portion of the interatrial septum has an abnormal horizontal orientation.

Chapter 8

Pulmonary Outflow Obstruction, Intact Septum

Pulmonary Outflow Obstruction, Intact Septum

- Pulmonary outflow tract obstruction can occur below, at the level of, or above the pulmomry valve.
- Each of these may occur in isolation or in association with other levels of pulmonary outflow obstruction in the same patient.

Pulmonary Valvar Stenosis
Double-Chambered RV
Supravalvar Pulmonary Stenosis
Pulmonary Atresia with Intact Ventricular Septum (IVS)

Valvar Pulmonary Stenosis

- Valvular pulmonary stenosis is characterized by fused valve leaflets which may be thickened or dysplastic.
- The valve anulus may be hypoplastic, and the valve itself may be bicuspid.
- Secondary changes include post-stenotic dilation of the main pulmonary artery and right ventricular hypertrophy.

Echo Tips:

† Best shown from parasternal short axis view and subcostal views.

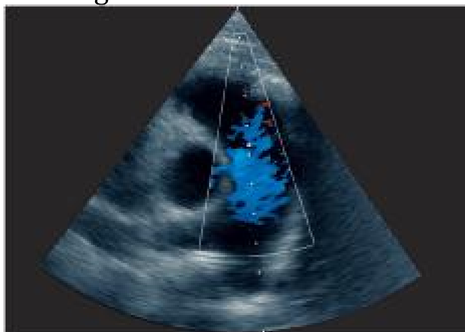
- The two-dimensional echocardiogram clearly demonstrates the typical features of the stenotic pulmonary valve from the standard and high parasternal short-axis and long-axis views as well as the subcostal sagittal views (Fig.below).
- The valve leaflets usually appear prominent because of thickening.
- Systolic motion is restricted, with inward curving of the tips of the leaflets, known as doming.
- Associated features, such as poststenotic dilation of the main and branch pulmonary arteries, also are easily recognized.
- Right ventricular hypertrophy, contractility of the right ventricle, as well as anatomy and function of the tricuspid valve should be assessed.
- Evidence of dynamic subpulmonary stenosis should be sought, but the severity may be impossible to estimate in the presence of more than mild valvar stenosis.
- The diagnosis of dysplastic pulmonary valve usually can be ascertained by echocardiography. The leaflets appear thickened and immobile, *without the characteristic doming seen in typical cases.* The pulmonary valve annulus is hypoplastic, and supra-

annular narrowing of the proximal main pulmonary artery is often present. *The poststenotic pulmonary artery dilation seen in classic cases is absent*.

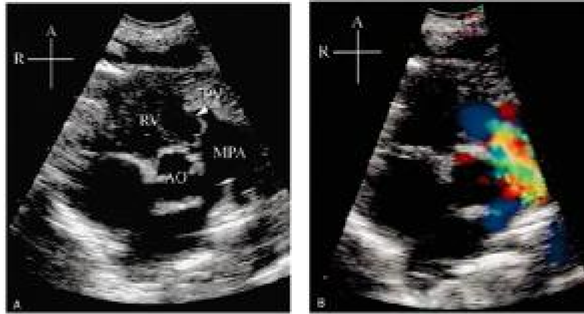
- The site of stenosis, anular size and presence of pulmonary or tricuspid regurgitation are important.
- The morphology and mobility of the valve leaflets must be assessed.
- The *severity of stenosis* should be assessed based on
  - measurement of right ventricular systolic pressure,
  - peak pressure gradient,
  - anular size,
  - right ventricular hypertrophy and function,
  - and the direction of shunting at atrial level.

Doppler Evaluation:

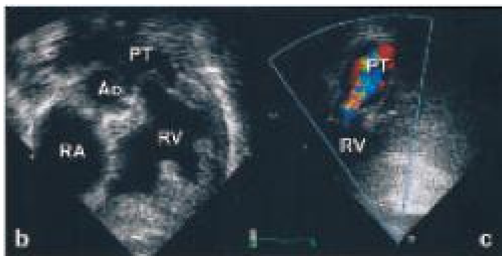
- The Doppler echocardiogram allows quantitative assessment of severity of pulmonary valve stenosis by estimating the pressure drop across the pulmonary valve (Fig. 40.9).
- The simplified Bernoulli equation $P = 4V_2^2$ is used, where P is the *peak* instantaneous *pressure* gradient, in millimeters of mercury, across the obstructed pulmonary valve, and V_2 is the *peak flow velocity*, in meters per second, distal to the obstructive orifice.
- If significant subpulmonary stenosis coexists, V_1 (the peak flow velocity proximal to the obstruction) must be taken into account.
- The echo beam must be aligned parallel with the main pulmonary artery trunk or the direction of the flow jet as seen on color Doppler.
- If tricuspid insufficiency is present, the Doppler technique can be used to calculate the pressure difference (P) between the right atrium and right ventricle by measuring the *peak flow velocity (V) of the tricuspid insufficiency jet*. Right ventricular pressure(Pulmonary Artary Pressure) then can be estimated by adding the pressure gradient to the estimated right atrial pressure.
- Color Doppler two-dimensional echocardiography has contributed to the diagnostic accuracy of pulmonary valve stenosis by demonstrating an abnormal flow pattern originating at the stenotic valve (Fig. below).
- *Normal flow is coded as red or blue*(in general), depending on whether it is directed toward or away from the transducer, respectively.
- High-velocity, turbulent flow through stenotic lesions appears as a mosaic jet with green, yellow, and other shades.
- Visualization of the jet by color also facilitates optimization of the alignment between the Doppler sample volume and the direction of flow, increasing the accuracy of the measured gradient.



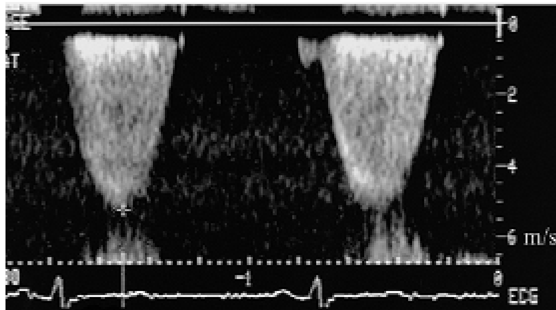
Normal pulmonary flow(Parasternal short-axis view)



Parasternal short-axis view from a 1-day-old infant with critical pulmonary valve stenosis . Note the thickened, doming leaflets in systole. The pulmonary valve (PV) annulus is smaller when compared with the aortic valve annulus. There is poststenotic dilation of the main pulmonary artery (MPA). B: Color Doppler through the stenotic pulmonary valve shows a mosaic pattern indicating high-velocity, turbulent flow originating at the level of the pulmonary valve. A, anterior; AO, aorta; R, right; RV, right ventricle.



(b) *Subcostal right oblique section in critical pulmonary valve stenosis shows a dome-shaped and dysplastic valve.*
(c) Parasternal anterior section with a narrow colour flow jet passing through the pulmonary valve. There is severe right ventricular
hypertrophy.



Continuous-wave Doppler recording from the parasternal short-axis view of a patient with severe pulmonary valve stenosis. The high-velocity systolic flow with a negative deflection from the baseline results from a jet directed away from the transducer from the right ventricle to the main pulmonary artery. The peak velocity of the jet is 5.2 m/s, equivalent to a peak pressure gradient of 110 mm Hg across the pulmonary valve.

Postop:

- The treatment of choice for valvar pulmonary stenosis is balloon valvuloplasty.
- Patients with dysplastic pulmonary valves may not respond well to this modality and may require surgical valvotomy, occasionally with a transanularpatch.
- Post-intervention or postoperative echocardiograms should assess for residual pulmonary outflow obstruction, noting that sub-valvar obstruction maybe unmasked following relief of valvar stenosis.
- Right ventricular pressure and function, and the presence and degree of pulmonary and tricuspid valve insufficiency should be assessed.

Double-Chambered Right Ventricle

- Hypertrophy of the septal band, moderator band and parietal band of the right ventricle cause stenosis of the proximal os infundibuli, dividing the right ventricle into a proximal, high-pressure, muscle-bound chamber, and a distal, low-pressure, smooth-walled infundibulum.

Echo Tips:

- Best shown from subcostal views.
- The site and mechanism of obstruction should be identified.
- Additional levels of obstruction should be sought.
- *Subaortic membranes* are a common associated finding, and should be ruled out.

Postop:

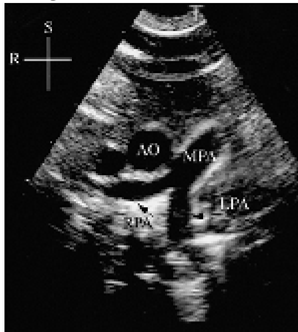
- Surgery consists of resection of obstructive right ventricular muscle and correction of associated pathology such as ventricular septal defects.
- Postoperative echocardiograms should assess for residual outflow tract obstruction. All levels of the pulmonary outflow tract should be assessed since distal obstruction may be unmasked after the relief of proximal obstruction.

Supravalvar Pulmonary Stenosis

- May occur in isolation or in association with ventricular septal defects or valvar pulmonary stenosis.
- *Commonly seen as part of congenital rubella syndrome*. In moderate or severe cases, secondary right ventricular hypertrophy may result.

Echo Tips:

- Best show n from parasternal short axis.
- The anatomy of the proximal pulmonary arteries usually can be delineated fairly well by echocardiography, but the distal pulmonary arteries cannot be imaged reliably.
- The parasternal short axis view and suprasternal views are the most helpful (Fig. below).
- Color flow Doppler can contribute to the qualitative assessment of stenosis by the appearance of turbulence at the area of obstruction.
- The pressure gradient can be estimated by Doppler but not always accurately.
- The echocardiogram is useful in detecting *secondary manifestations of right ventricular hypertension*, such as right ventricular hypertrophy, tricuspid insufficiency, or enlargement of the right-sided chambers.
- The obstructive segment should be located and measured.
- Additional levels of obstruction should be sought.



Suprasternal notch short-axis view from an infant with Williams syndrome and mild proximal right pulmonary artery (RPA) and left pulmonary artery (LPA) stenosis. The proximal branches are well seen, but the peripheral branches cannot be well imaged. AO, aorta; MPA, main pulmonary artery; R, right; S, superior.

Postop:

- Treatment of supravalvepulmonary stenosis is surgical resection.
- The results of surgery are dependent on the state of the distal pulmonary arterial tree; multiple levels of peripheral pulmonary stenosis are associated with poor outcomes.
- Postoperative echocardiograms should include careful assessment of the entire pulmonary outflow tract up to the hilum of both lungs, looking for multilevel outflow obstruction and distal branch pulmonary artery stenosis.
- *Echocardiography is insensitive for detection and quantification of branch pulmonary artery stenosis distal to the pulmonary hilum*.

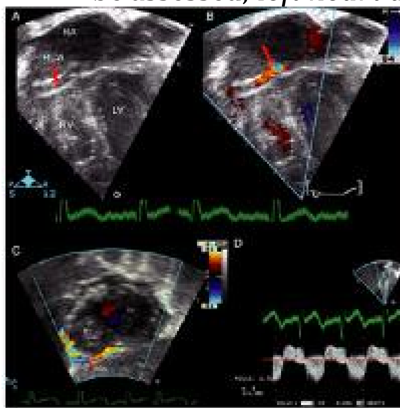
Pulmonary Atresia, Intact Ventricular Septum

- This lesion represents the extreme end of the spectrum of pulmonary valve stenosis with an intact ventricular septum.
- Absence of prograde flow across the pulmonary valve leads to variable degrees of hypoplasia of the light ventricle and tricuspid valve.
- The right ventricle continues to attempt to eject against the imperforate pulmonary valve, resulting in high (even suprasystemic) right ventricular pressure; right ventricular hypertrophy and tricuspid regurgitation may result.
- High right ventricular pressure may lead to right ventricular sinusoids which connect with the coronary arteries (ventriculocoronary connections) with stenosis or absence of the proximal aortocoronary connections.
- As a result, part or all of the coronary circulation may be dependent on the right ventricle.

Echo Tips:

- Best shown from apical four chamber and parasternal short axis.
- The role of echocardiography is to
  - diagnose this condition,
  - to assess the size and function of both ventricles and the tricuspid valve,
  - and to determine the patency of the ductus arteriosus which maybe the only source of pulmonary blood flow until palliative shunt surgery.
  - *Angiography is needed to establish the details of coronary artery anatomy*.
- Because of the obligatory right-to-left shunt at atrial level, with rare exception, there is either a patent foramen ovale or true secundum atrial septal defect.
- The tricuspid valve rarely is normal in patients with pulmonary atresia and intact ventricular septum.
- Aortic valve stenosis has been described in patients with pulmonary atresia and intact ventricular septum, including the neonate with critical aortic stenosis and the somewhat older child with severe aortic valve stenosis.
- The myocardium of patients with pulmonary atresia and intact ventricular septum can demonstrate a wide range of abnormalities. Ischemia, fibrosis, infarction, and myocardial rupture have been observed in these patients
- The size of the right ventricle correlates with the diameter of the tricuspid anulus, which should be measured and converted to a Z-score (normalized to body surface area) using available nomograms.
- The diameters of the main and branch pulmonary arteries should be measured.

- All potential sources of pulmonary blood flow including the ductus arteriosus should be assessed.
- The presence of ventriculocoronory connections may be suspected *based on low velocity flow within the right ventricular myocardium*, shown by adjusting color flow Doppler to a low Nyquist limit.
- Other abnormalities include myocardial disarray, the appearance of so-called spongy myocardium, and ventricular endocardial fibroelastosis.
- *Right ventricular systolic pressure* should be quantified *using the peak velocity of tricuspid regurgitation*. The severity of tricuspid regurgitation and the degree of right atrial dilation should also be assessed.
- Since all of the systemic venous return must cross the atrial septum, the size and adequacy of interatrial communication must be assessed.
- Left ventricular size and function and the presence/ severity of mitral regurgitation should be assessed; *left heart dysfunction maybe due to coronary insufficiency*.



Four-chamber apical view depicting a dilated right coronary artery in the atrioventricular groove. B: Color flow Doppler of the same image confirms flow within the vessel. C: Subcostal imaging of the epicardial right coronary artery confirms dilation to suggest increased flow. D: Pulsed Doppler confirms systolic retrograde flow with diastolic antegrade flow in the coronary vessel with connections to the hypertensive right ventricle. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle; red arrows, dilated RCA (right coronary artery).

Postop:

- Surgical options range from single ventricle staged repair for patients in whom the right ventricle is not salavageable (go to Chapter 31), to the need for cardiac transplantation for patients who have severe coronary arterial abnormalities or occlusion (go to Chapter 30).
- Biventricular repair is feasible inpatients with adequate right ventricular and tricuspid valvar size and function. This is usually a staged procedure, involving a combination of aortopuunonary shunt operations and pulmonary outflow tract reconstruction establishing prograde flow from the right ventricle to the main pulmonary artery.
- Echocardiograms obtained during staged biventricular repair should assess for tricuspid anulus size and biventricular size and function. *The diameters of the branch pulmonary arteries, right ventricular systolic pressure, amount and direction of flow across the atrial septum should all be assessed*. The patency of aortopulmonary shunts that may have been placed should also be assessed.

Chapter 9

Tetralogy of Fallot

Tetralogy of Fallot

- This conotruncal malformation is the *most common congenital cyanotic heart defect*.
- The constellation of features that comprise tetralogy is attributed to *anterior, superior and leftwards malalignment* of the conal septum, which results in a large ventricular septal defect that is committed to the right ventricular inflow and outflow tracts.
- The deviated conal septum raises the floor of the right ventricular outflow tract, leading to pulmonary outflow tract obstruction(*the most characteristic and hallmark finding is the subpulmonic stenosis created by the deviation of the outlet, or conal, septum*)
- Malalignment of the conal septum carries the aorta with it, thus leading to an overriding aorta.
- Varying degree of conal septal malalignment lead to varying degrees of pulmonary outflow tract obstruction. Since flow creates growth, varying degrees of under development of the pulmonary outflow tract, anulus and branchpulmonaiy arteries may be seen.
- The significance of aortic override primarily relates to terminology and in distinguishing whether the anatomic entity in question is more appropriately deemed to be *double-outlet right ventricle or TOF*. This issue may be avoided if the morphologic definition of double-outlet right ventricle is adopted. In this approach, *double-outlet right ventricle denotes the absence of aortic-mitral continuity and requires the presence of both a subaortic and subpulmonic muscular conus*.
- The aortic arch is right sided in TOF in approximately 25% of cases.

Classic
With Pulmonary Atresia
With Absent Pulmonary Valve

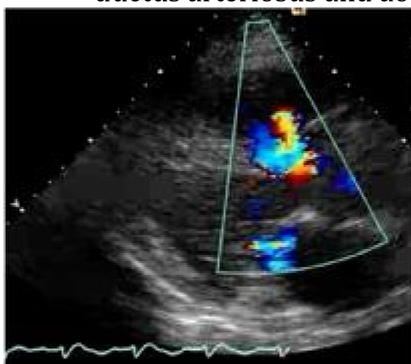
Classic Tetralogy of Fallot

- The classic form of tetralogy of Fallot is characterized by anterior, superior and leftwards malalignment of the conal septum leading to subpulmonary stenosis, yet maintaining prograde pulmonary blood flow, a large malalignment-type ventricular septal defect and an overriding aorta.
- Varying degrees of conal septal malalignment and hypertrophy lead to subpulmonary stenosis of varying severity.
- The pulmonary anulus may be hypoplastic; valve leaflets maybe thickened, and comnissural underdevelopment may lead to a bicuspid pulmonary valve.
- In the presence of prograde pulmonary blood flow, the branch pulmonary artery diameters are usually within normal range, and aortopulmonary collaterals are uncommon. This is dramatically different from the scenario of tetralogy of Fallot with pulmonary atresia, which is discussed separately in the following section.

Echo tips:

- Anterior, leftward and superior deviation of the conal septum is best shown from *subcostal short axis* view with anterior angulation and clockwise rotation of the transducer.

§ The right ventricular outflow tract is also well profiled from *parasternal short axis views*.
§ Aortic override is best shown from parasternal long axis view.
§ Right ventricular outflow obstruction can occur at multiple levels:
ü sub -PS due to deviation of the conal septum and muscular hypertrophy,
ü valvar PS due to anular hypoplasia and/or fused, dysplastic leaflets,
ü supravalvar PS and branch puhmonary artery hypoplasia or stenosis can occur.
Each of these levels should be specifically interrogated.
§ Additional VSD's should be sought.
§ Since right ventricular pressure is usually elevated, the color sweep of the ventricular septum should utilize a *low Nyquist limit*, The direction of VSD flow should be shown with pulsed wave and color flow Doppler.
§ Additional sources of pulmonary blood flow, such as aortapulmonary collaterals, patent ductus arteriosus and aortopulmonary window must be ruled out.



Color doppler left parasternal long axis view shows an over-riding aorta with ventricular septal defect and left-to-right shunt.



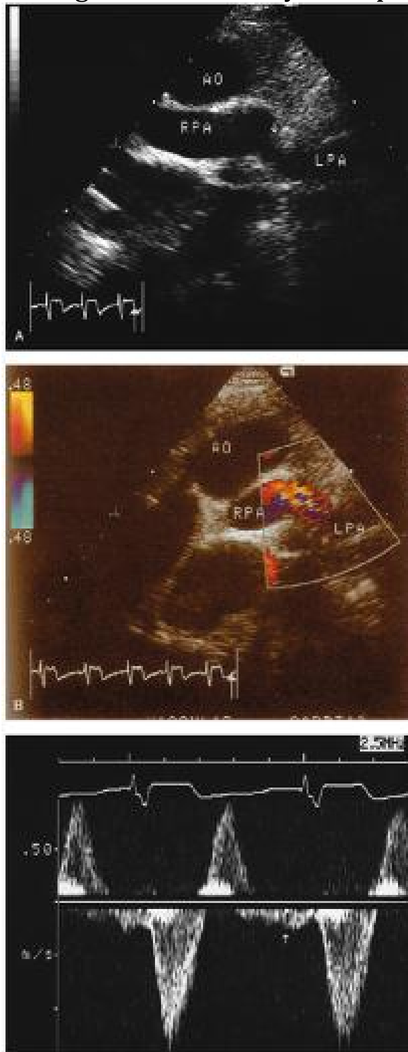
This subcostal two-dimensional echocardiographic view demonstrates the anterior deviation of the conal septum (arrow) and the ventricular septal defect. CS, conal septum; INF, infundibulum; LV, left ventricle; PT, pulmonary trunk; RV, right ventricle.

Tetralogy of Fallot – Postop:

§ Surgery for tetralogy of FaIlot maybe either a
ü staged repair, with an aortopulmonary shunt followed by shunt take-down and complete repair,
ü or a singlestage correction .Most commonly, a modified Blaloc k-Taussig (B-T) shunt is used.
§ *A Modified Blaloc K-Taussig (B-T) Shunt* :This is a Goretex tube connecting the side of the subclavian artery to the side of the ipsilateral branch pulmonary artery. This shunt is usually

placed on the side opposite the aortic arch; thus, in a patient with a left aortic arch, the shunt would be placed on the right side.

- Echocardiograms obtained following shunt surgery should assess for shunt patency and caliber along the entire length of the shunt .
- The branch pulmonary arteries should be assessed for distortion or stenosis in the vicinity of the shunt insertion.
- *Complete repair of tetralogy of Fallot* consists of patch closure of the ventricular septal defect, and augmentation of the right verntricular outflow tract .Depending on the size of the pulmonary anulus, trsnsanular patch may be needed. In addition, a pulmonary valvectomy is usually performed. Some centers place a pericardial monocusp valve inaplacec of the pulmonary valve in order to minimize pulmonary insufficiency.
- Echocardiograms obtained following complete repair of tetralogy of Fallot should assess :
  - for residual VSD and right ventricular outflow tract obstruction.
  - The branch pulmonary arteries should be measured.
  - The severity of pulmonary insufficiency should be quantified.
  - Right ventricular systolic pressurr should be estimated.



This high parasternal short-axis two-dimensional echocardiogram demonstrates left pulmonary artery narrowing (arrow) in a 10-year-old repaired at 6 months of age. B: Color flow mapping (red) shows pulmonary regurgitation arising from the left pulmonary artery. C: Pulsed Doppler in the main pulmonary artery shows low-velocity antegrade

flow. Retrograde diastolic flow occurs only through one third of diastolic and is followed by antegrade diastolic flow with atrial systole (arrow). AO, aorta; LPA, left pulmonary artery; RPA, right pulmonary artery.

Tetralogy of Fallot with Pulmonary Atresia

- This condition is due to extreme malalignment of the conal septum, leaving either a slit like subpulmonary infundibulum or, if the conal septum fuses with the free wall of the right ventricular outflow tract, infundibular atresia.
- This seals off the right ventricular outflow tract, preventing prograde flow into the pulmonary circulation and resulting in varying degrees of hypoplasia of the branch pulmonary arteries.
- Pulmonary blood flow maybe provided by various additional sources including the ductus arteriosus and aortopulmonary collaterals.
- Echocardiography serves as a diagnostic tool, but usually does not establish the comprehensive diagnosis in this condition. Angiography and/or MRI studies are needed to define the extent, size and distribution of the various sources of pulmonary blood flow.

Echo Tips:

- The branch pulmonary arteries are frequently diminutive, and may not be confluent. Pulmonary arterial anatomy must be defined in each case.
- The smaller the branch pulmonary arteries, the higher the probability of collateral arterial supply from the aorta to the lungs. It is difficult to define dime collaterals completely by echocardiography;however, the origin of these collaterals from the descending thoracic aorta and the brachiocephalic vessels can be defined.
- A long, tortuous 'curly-cue' patent ductus arteriosus is frequently present.

Tetralogy of Fallot, Pulmonary Atresia – Postop:

- Surgery for tetralogy of FaIlot with pulmonary atresia may be either a staged repair , with an aortopulmonary shunt followed by shunt take-down and complete repair, or single-stage correction. The presence of aortopulmonary collaterals may complicate the situation, necessitating incorporation into the repair (unifocalization) or coil embolization.
- Most commonly, modified Blabck-Taussing (B-T) shunt is used.
- An alternative approach is to place a conduit between the right ventricle and the main pulmonary artery (leaving the ventricular outlet open) to establish prograde flow into the pulmonary arteries to encourage pulmonary arterial growth.
- Echocardiograms obtained following staging surgery should assess for shunt conduit patency and caliber along the entire length of the shunt conduit .The branch pulmonary arteries should be assessed for distortion or stenosis in the vicinty of the shunt insertion.
- *Complete repair* consists of patch closure of the ventricular septal defect to the aorta (*Rastelli procedure*), and usually, placement of a conduit (typically s pulmonary homograft) between the right ventricle and the pulmonary arterial bifurcation.
- Echocardiograms obtained following complete repair should
  - assess for residual VSD and right ventricular outflow tract obstruction
  - The branchpulmonary arteries should be measured.
  - The severity of pulmonary insufficiency should be quantified.
  - Right ventricular systolic pressure should be estimated.

Tetralogy Of Fallot With Absent Pulmonary Valve Syndrome

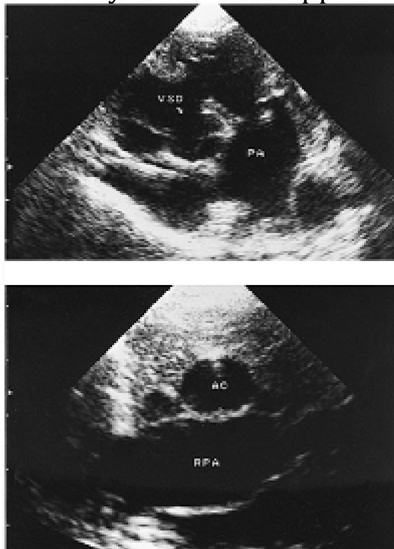
- anatomic findings consist of a incompletely formed, rudimentary pulmonary valve that typically is both stenotic and regurgitant, aneurysmally dilated pulmonary arteries, and a

large malaligned outlet ventricular septal defect (VSD). The association of the VSD with this disorder is nearly uniform.

- This syndrome also is virtually always associated with absence of the patent ductus arteriosus.
- Other clinical features fairly typical in this disorder include the common association of airway abnormalities that may lead to severe respiratory failure.
- In addition to the typical features of tetralogy of Fallot (enumerated earlier in this chapter), this unusual anomaly is characterized by absence of functional pulmonary valve tissue.
- Rudimentary tags of tissue are frequently present at the anulus however these are not enough to allow leaflet coaptation and valve closure. The result is severe pulmonary insufficiency.
- While the conal septal malalignment seen in this anomaly is identical to that seen in other cases of tetralogy of Fallot, the dominant hemodynamic lesion is pulmonary insufficiency leading to right ventricular dilatation.
- As a result of this dilatation, the right ventricular outflow tract is not stenotic.
- The branch pulmonary arteries are aneurysmally dilated in this condition; this dilation may extend into the parenchyma of the lung, leading to compression of the airway and parenchyma.
- Pulmonary arterial dilation is frequently progressive even after surgical repair, mandating periodic echocardiographic assessment of the branch pulmonary arteries. It is speculated that absence of the ductus arteriosus leads to this condition.

Ech Tips:

- The pulmonary valve is best shown from parasternal short axis;
- rudimentary tags of valve tissue are frequently seen and the valve leaflets usually do not coapt.
- Patent ductus arteriosus is rarely seen with this lesion.
- Aneurysmal dilatation Of the branch pulmonary arteries is very common; these arteries should be measured for purposes of serial assessment.
- Severe, low velocity pulmonary insufficiency is the rule; the width of the jet is best assessed by color flow Doppler with a low Nyquist limit.



Two-dimensional echocardiogram in a neonate with absent pulmonary valve syndrome. The short-axis view (upper panel) shows a ventricular septal defect (VSD) and a large main pulmonary artery (PA). Note that the pulmonary valve leaflets are rudimentary but not actually absent. The suprasternal notch image (lower panel) shows a markedly dilated right pulmonary artery (RPA) passing beneath the aortic arch (AO).

Tetralogy of Fallot, Absent Pulmonary Valve – Postop:

- Surgery for tetralogy of Fallot with absent pulmonary valve is usually a single-stage correction This consists of patch closure of the ventricular septal defect, and augmentation of the right ventricular outflow tract .Depending on the size of the pulmonary anulus, a transanular patch maybe needed. In addition, a pulmonary valvectomy is usually performed. Either a pericardial monocusp valve or a pulmonary homograft conduit is placed in the pulmonary position. The aneurysmaIly ditated pulmonary arteries may need to be resected to normal size.
- Echocardiograms obtained following compIete repair of tetralogy of Fallot should assess for residual VSD and right ventricular outflow tract obtruction The branch pulmonary arteries should be measured serially, looking for progressive dilation The severity of pulmonary insufficiency should be quantified. Right veniricular systolic pressure should be estimated.

Chapter 10

The Pulmonary Arteries

The Pulmonary Arteries

- The main pulmonary artery is separated from the aorta by the aortopulmonary septum.
- The branch pulmonary arteries are formed from the developing lung buds and fuse with the main pulmonary artery *to create the normal pulmonary arterial confluence*.
- The main pulmonary artery courses *posteriorly*, *superiorly and leftwards* from its origin, and the left pulmonary artery follows this orientation .
- The right pulmonary artery arises at an angle from the main pulmonary artery, and courses posterior to the ascending aorta and the main pulmonary artery.

This chapter illustrates several pulmonary arterial anomalies that are encountered in children with congenital heart disease.

Focal Stenosis
Diffuse PA Hypoplasia
Criss-cross Arrangement
Absent RPA
Branch PA Dilation

Focal Stenosis

- Focal stenosis of the branch pulmonary arteries maybe a native or a post-operative phenomenon. This is commonly seen inpatients with decreased pulmonary blood flow, eg., Tetralogy of Fallot especially with pulmonary atresia, or d-transposition of the great arteries with pulmonary stenosis.
- Stenosis may occur proximally, at the origin of the branch pulmonary arteries.
- In patients who have undergone aor to-pulmonary shunts as palliation for cyanotic heart disease, acquired stenosis of the branch pulmonary arteries may be seen in the vicinity of the site of shunt insertion due to 'clamp injury'.

Diffuse Hypoplasia

- Diffuse hypoplasia of the branch pulmonary arteries is *classically seen in patients with tetralogy of Fallot and pulmonary atresia*.
- There may be multiple sources of pulmonary blood flow, including the ductus arteriosus and aortopulmonary collaterals.
- The pulmonary arteries may be nonconfluent or discontinuous in this scenario.
- The complete diagnosis of this condition, including details of collateral blood flow to the lung segments, uually requires cardiac catheterization and/or MRI studies.

ECHO TIP:

- The pulmonary arterial confluence is best shown from subcostal long axis view.
- The right pulmonary artery is best shown from suprasternal short axis views, and parasternal short axis view with clockwise rotation of the transducer into a more transverse (right-left) orientation.

- The left pulmonary artery is best shown from suprasternal long axis views, and parasternal short axis with counter-clockwise rotation of the transducer into a more antero-posterior plane.
- The site and severity of hypoplasia must be shown with 2-D imaging using high-frequency transducers.
- Measurements of the pulmonary arteries must be made in more than one plane, and confirmed with color flow Doppler (measuring the diameter of the flow jet).
- Doppler gradients maybe underestimated in the presence of proximal obstruction, such as valvar pulmonary stenosis, or elevated distal pressures, as in a neonate or with pulmonary hypertension, and also due to the long, tunnel-like nature of pulmonary arterial hypoplasia.
- Right ventricular pressure must be estimated using the peak velocity of tricuspid regurgitation.
- Additional sources of pulmonary blood flow, such as a ductus arteriosus and aortopulmonary collaterals should be sought .It must be determined whether prograde flow from the ventricles into the pulmonary arteries exists, and whether the pulmonary arteries are confluent.
- Currently, echocardiography cannot reliably assess the pulmonary arteries beyond the hilum of the lung; other imaging modalities are needed to clarify the issues of intrapulmonary hypoplasia of the branch pulmonary arteries, and the distribution of collateral blood flow to the lungs.

Criss-Cross Arrangement

- So-called 'criss-cross' arrangement of the pulmonary arteries refers to an unusual arrangement of the origins of the branch pulmonary arteries.
- In this anomaly, the left pulmonary artery originates from the rightwards, anterior aspect of the main pulmonary artery.
- The right pulmonary artery originates from the leftwards, posterior aspect of the main pulmonary artery, and courses behind the main pulmonary artery to reach the right lung.
- This leads to an unusual echocardiographic appearance but does not have any hemodynamic significance.
- This anomaly maybe associated with a structurally normal heart, or may occur with complex heart disease; eg. in a patient with interrupted aortic arch type B.

ECHO TIP:

- Best shown with a right to left sweep from the high left parasternal ('ductal') view.
- Focal areas of pulmonary artery stenosis must be ruled out.
- Neither the trachea/ mainstembronchi nor the esophagus are associated with this anomaly; therefore, this anomaly does not cause a vascular ring or vise.

Absent Right Pulmonary Artery

- If the right pulmonary artery is completely absent, the right lung is supplied by aortopulmonary collaterals.
- While echocardiography may not identify the right pulmonary artery, magnetic resonance imaging usually shows a small remnant at the Mum of the right lung.
- This anomaly may occur with a structurally normal heart .
- It may be impossible to achieve complete echocardiographic definition of the aortopulmonary collaterals that supply the right lung. Realistically, the role of

echocardiography is detection of this lesion; complete definition of the diagnosis usually requires cardiac catheterization, and/or MRI studies.

ECHO TIP:

- Best shown from suprasternal long axis view which shows absence of the right pulmonary artery, usually seen in cross-section immediately below the aortic arch.
- Also shown from parasternal short axis view and subcostal long axis view which shows the main pulmonary artery connecting only to the left pulmonary artery, with no identifiable right pulmonary artery.
- An attempt must be made to identify the sources of blood flow to the right lung, whether via collaterals from the aorta or via the ductus arteriosus.
- Additional tests, such as angiography and MRI studies, are usually needed for a comprehensive diagnosis in this condition.

Dilated Branch Pulmonary Arteries

- Diffusely dilated branch pulmonary arteries are classically associated with tetralogy of Fallot with absent pulmonary valve and severe pulmonary insufficiency.
- While echocardiographic imaging of the pulmonary arteries is not adequate distal to the hilum, these patients frequently exhibit dilation of the intraparenchymal pulmonary arterialbed, leading to serious long-term respiratory pathology from compression of the airways and lung parenchyma.
- For more details on tetralogy of Fallot with absent pulmonary valve syndrome, go to Chapter 9.

ECHO TIP:

- Best shown from suprasternal views.
- The diameters of the the pulmonary arteries must be measured at multiple sites for serial comparison.
- Any localized dilation (aneurysm) of the branch pulmonary arteries must be identiied and quantified.

Chapter 11

Mitral Valve and Left Atrium

Mitral Valve & Left Atrium

- The left atrium and mitral valve develop from :
  - the common pulmonary vein,
  - the posterior waII of the developing atrial chamber,
  - septum primum,
  - the atrioventricular canal septum
  - and the left atrial appendage (primitive atrial tissue).
  - The subvalvar apparatus of the mitral valve develops from left ventricular myocardium.
- The function of the left atrium and mitral valve is to provide unobstructed and non-regurgitant flow from the pulmonary veins into the left ventricle. This, in turn, requires normal fusion of the left atrium with the common pulmonary vein, and normal development of the left atrial cavity, mitral anulus, leaflets, chordae tendineae and papillary muscles.
- Defective development of any of these components can lead to congenital anomalies involing the left atrium and the mitral valve and affecting left ventricular inflow.

Mitral Valve Prolapse
Cleft Mitral Valve Leaflet
Mitral Stenosis
Mitral Regurgitation
Double Orifice Mitral Valve
Cor Triatriatum
Supramitral Ring

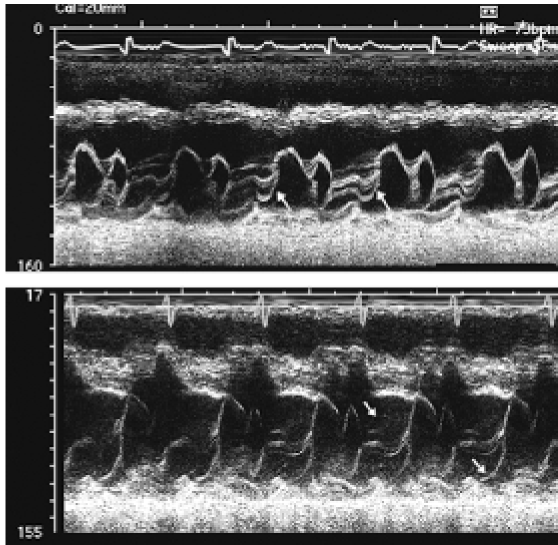
Mitral Valve Prolapse

- The incidence of mitral valve prolapse depends on the definition of the condition.
- There are two basic forms of mitral valve prolapse that represent the two ends of a spectrum of abnormality.
  - The first, which represents a true form of organic heart disease, is mitral valve prolapse associated with myxomatous thickening of the mitral valve leaflets.
  - The second form of mitral valve prolapse represents mild buckling of an otherwise anatomically normal valve.
- In general terms, this refers to movement of the mitral valve leaflets (most commonly the anterior mitral leaflet) posteriorly and superiorly beyond the plane of the mitral anulus during ventricular systole.
- The parasternal long axis view has been classically used for establishing or ruling out mitral valve prolapse.
- However, it has been pointed out that the plane of the mitral anulus is not flat throughout ventricular systole, and that *use of a single plane to diagnose mitral valve prolapse may lead to a high Incidence of this condition*.

- True mitral valve prolapse is frequently seen inpatients with Marfan syndrome. Here, the condition is due to elongated, redundant chordae tendineae.
- The mitral valve leaflets are also frequently abnormally thickened, scalloped and redundant.
- Mitral regurgitation may be an associated finding, and is due to non-coaptation of mitral valve leaflets *w*hich is increasingly manifest as systole progresses, leading to increasing tensile stretch on the chordae tendineae. Thus, mitral regurgitation may be seen in mid-systole or late systole.
- Isolated MVP is a rare cause of mitral insufficiency in the first year of life, and although the cause of the condition is unclear, neither clinical features nor two-dimensional (2-D) echocardiographic findings of mitral valve prolapse have been seen in newborns . Thus, the phenotypic expression of prolapse becomes more frequent with advancing age.

ECHO TIP:

- Best shown from parasternal long axis view; other views should be used to confirm the diagnosis.
- With M-mode echocardiography), mitral valve prolapsed was diagnosed in the presence of leaflet thickening with posterior bowing of the mitral valve apparatus during systole. This bowing could be present either throughout entire systole or confined to late systole.From a technique standpoint, it is important to recognize that the M-mode interrogation beam should be aligned in a manner to encompass the area just behind the mitral anulus if one is to document buckling of the mitral valve leaflet into the left atrium.
- Two-dimensional echocardiography is a more commonly employed technique for screening for mitral valve prolapsed:
  - It is more important to appreciate the presence or absence of valve thickening and the symmetry versus asymmetry with which the valve was prolapses.
  - As a general rule, the *four-chamber view will be less specific* for the diagnosis of mitral valve prolapsed than detection of buckling in either a parasternal long-axis view or apical two-chamber view.
- Each component of the mitral valve - the anulus (size), each leaflet (prolapse, redundancy, coaptation), chordae (redundancy, rupture leading to a flail leaflet), and papillary muscles (function) - should be assessed systematically.
- The presence, timing (pansystolic, mid-systolic or late systolic) and severity of mitral regurgitation should be assessed. The location and the width of the regurgitant jet should be defined from a parasternal short axis sweep using color flow Doppler.
- Additional information - including left atrial and left ventricular' size and presence or absence of systolic flow reversal in the pulmonary veins - enables quantification of the severity of mitral regurgitation.
- The diagnosis of mitral valve prolapse should be made when one or both leaflets breaks the plane of the mitral anulus in a nonsymmetric manner, typically taking on a buckling appearance. As noted previously, the leaflet should be described as thickened or anatomically normal as well.
- Patients with mitral valve prolapse may have Marfan syndrome, with associated aortic root dilation which maybe progressive; serial measurements of aortic root and sino tubular junction diameter must be performed.



M-mode echocardiograms recorded in two patients with mitral valve prolapse. In each instance, note the distinct posterior motion of the mitral valve (arrow). Bottom: Note the chordal systolic anterior motion (upper arrow), which may also be seen in mitral valve prolapse.

Cleft Mitral Valve Leaflet

- This rare defect of the anterior mitral valve leaflet often is associated with significant mitral insufficiency presenting in infancy or young children, but it may occur as an isolated defect in asymptomatic individuals ; the atrioventricular septum is intact and left ventricular outflow tract not elongated.
- The cleft variably divides the anterior leaflet into more or less two components.
- The cleft may be partial within the leaflet or extend to the base of the valve.
- The valve cleft edges are thickened and rolled.
- The cleft is directed anteriorly toward the outflow septum or aortic root *in contrast to the cleft in atrioventricular septal defect* . Here the cleft is more posteriorly directed toward the inlet septum and located in a small common leaflet that bridges the septum.
- In isolated cleft of the mitral valve, the papillary muscles are variable but generally normal, and in most cases chordae attach to the papillary muscles, whereas accessory cords usually attach to the membranous and muscular septum. In some cases of complete cleft, accessory cords are absent, and the anterior leaflet is usually flail and grossly insufficient.
- Associated left ventricular outflow tract obstruction *may be caused by the accessory cords*, and associated congenital defects including ventricular septal defect are common.
- These features influence clinical findings and the time of presentation. The annulus is commonly dilated.
- Less commonly, *the posterior leaflet is involved*, sometimes termed partial leaflet agenesis. The leaflet tissue is deficient leading to a cleft that extends from the edge to the annulus. Chordae are deficient to absent at the cleft margin. The most commonly associated congenital heart anomalies include atrial and ventricular septal defects and transposition of the great arteries.

Mitral Stenosis

- Mitral stenosis may be an isolated anomaly, or may occur in association with other forms of left-sided obstructive lesions, including supramitral ring, aortic outflow tract obstruction,

coarctation of the aorta or as apart of hypoplastic left heart syndrome. It may also be a sequel of rheumatic valvulitis.

- This lesion may involve the anulus, leaflets, chordae tendineae and/or papillary muscles.
- Thus, the anulus may be hypoplastic; leaflets may be thickened and rolled with fused commissures; chordae maybe foreshortened or absent leading to direct insertion of the valve leaflets onto the papillary muscles; fused chordae may lead to obliteration of interchordal spaces.
- The papillary muscles may be fused or closely spaced, and the chordae of both leaflets may insert onto a single papillary muscle, leading to a parachute mitral valve.
- Any of these components may be seen in a given patient. The most severe end of the spectrum is mitral atresia, with no prograde flow across the mitral valve.
- Echocardiography should be able to establish the diagnosis comprehensively in most cases of mitral stenosis.

ECHO TIP:

- Ø Best shown from parasternal and apical views.
- Each component of the mitral valve must be assessed systematically.
- The mitral anulus must be measured in orthogonal views.
- Leaflet mobility, thickening or redundancy must be noted.
- The chordae tendineae should be assessed for foreshortening, fusion and adequacy of interchordal spaces.
- The number, spacing and location of the papillary muscles, and the pattern of chordal attachments to the papillary muscles (whether symmetric or predominantly to one papillary muscle) must be established using slow sweeps in the short axis of the ventricle.
- Additional mitral valve pathology, such as a supravalve ring, must be ruled out; for more details, see the section on Supramitral ring in this chapter.
- The presence and morphologic basis for associated mitral regurgitation must be identified.
- Quantitative Doppler echocardiography is useful, but may be difficult to apply to individual patients; the situation is complicated by high left atrial compliance which may lead to underestimation of the severity of stenosis.

POST OP:

- Either surgical or transcatheter (balloon) valvuloplasty may be performed to treat mitral stenosis.
- Post-intervention echocardiograms should assess for residual mitral stenosis and for the presence and severity of new or increased mitral regurgitation.
- A decrease in left atrial size is a useful indicator of the success of intervention.
- Pulmonary artery pressures should also be estimated using the *peak velocity of tricuspid regurgitation*.

Mitral Regurgitation

- Isolated congenital mitral regurgitation is very rare.
- Mitral regurgitation (= insufficiency) is most frequently associated with a structurally defective mitral valve, as with a cleft anterior leaflet.
- Other causes include mitral valve prolapse, ruptured mitral chordae leading to a flail leaflet, papillary muscle dysfunction due to ischemia, rheumatic valvulitis and hypertrophic cardiomyopathy.
- Echocardiography should be able to identify the morphologic basis and estimate the severity of mitral regurgitation

ECHO TIP:

- Ø Best shown from parasternal and apical views. The location and the width of the regurgitant jet should be defined from a parastemal short axis sweep using color flow Doppler.

The morphologic basis of mitral regurgitation should be defined.

- § Each component of the mitral valve - the anulus (dilation), each leaflet (prolapse, flail, redundancy, coaptation), the commissures (cleft), chordae (redundancy, rupture leading to a flail leaflet), and papillary muscles (function)- should be assessed systematically.
- § The timing (pansystolic, mid-systolic or late systolic) and severity of mitral regurgitation should be assessed.
- § Additional information - including left atrial and left ventricular size and presence or absence of systolic flow reversal in the pulmonary veins -enables quantification of the severity of mitral regurgitation.
- § Patients with mitral valve prolapse may have Marfan syndrome, with associated aortic root dilation which maybe progressive; serial measurements of aortic root and sino tubular junction diameter must be performed.

POST OP:

- § Treatment of mitral regurgitation depends on the cause and severity.
- § In patients with a severely dilated mitral anulus, surgery may consist of mitral anuloplasty with a ring placed at the level of the anulus to improve leaflet coaptation and prevent further anular dilation.
- § A cleft in the mitral leaflet maybe sutured closed; ruptured chordae maybe amenable to repair. In some cases, mitral valve repair maybe impossible, and valve replacement with a prosthesis may be needed.
- § Post-repair echocardiograms should assess for the presence, severity and location of residual mitral regurgitation. New or increased mitral stenosis may also be seen postoperatively, and should be sought specifically.
- § *A decrease in left atrial size and resolution of pulmonary hypertension (from preoperative levels) are good indicators of the adequacy of surgical intervention.*

Double Orifice Mitral Valve

- § This is a rare anomaly; it maybe associated with mitral stenosis or regurgitation.
- § It is characterized by a *tissue bridge* that extends from the anterior to the posterior mitral leaflet at the level of the anulus, thus dividing the mitral valve into two orifices. These orifices may be unequal in size; the chordal supporting structure may not be balanced, resulting in mitral stenosis and/or regurgitation.
- § This anomaly maybe associated with atrioventricular canal defects.

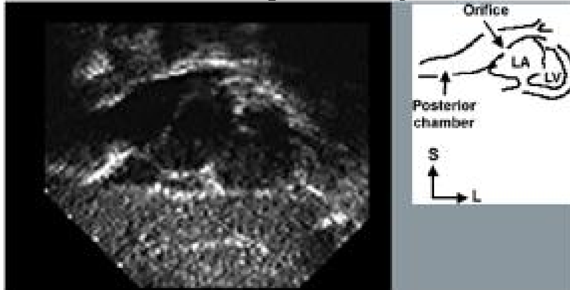
ECHO TIP:

- Ø Best shown from apical and parasternal short axis views.
- § Each of the orifices must be assessed for regurgitation.
- § Prograde flow profiles across the mitral valve should be assessed for evidence of stenosis.
- § If one of the orifices is regurgitant due to inadequate chordal support, then echocardiography should determine whether the other orifice is adequate to support cardiac output, since the only surgical option maybe closure of the regurgitant orifice.

Cor Triatriatum

- § Normal left atrial development depends on fusion between the embryonic common pulmonary vein with the confluence of the right and left pulmonary veins behind the primitive atrium.

- Following this fusion, both the common pulmonary vein and the venous confluence must be completely incorporated into the primitive atrium, giving rise to the smooth walled venous portion of the chamber.
- Cor triatriatum is due to incomplete incorporation of these embryonic structures into the primitive atrium. This results in the echocardiographic appearance of *two left-sided 'atrial chambers' separated by a membrane*.





Apical 4 chamber view of a non-obstructive membrane in the left atrium consistent with 'cor triatriatum'.

- The more *posteriorly*, *superiorly* located chamber receives the pulmonary veins. The true left atrium is located anterior and inferiorly, and communicates with the left atrial appendage, the mitral valve and the left ventricle.
- Cor tiatriatum presenting as a non-obstructing membrane in the left atrium.
- The communication between the posterior chamber and the true left atrium may be restrictive, resulting in pulmonary venous obstruction.
- In atypical forms, the posterior chamber may communicate with either the right atrium or with systemic veins.

ECHO TIP:

- Best shown from subcostal long axis and apical views.
- Seen as an echo dense *curvilinear shadow* behind the left atrium, protruding towards the mitral valve.
- The membrane is located posterior and superior to the left atrial appendage and the foramen ovale and moves towards the mitral valve in ventricular diastole.
- Each of the pulmonary veins should be identified and tracked into the posterior clamber. Any connections of this chamber with either the right atrium or systemic veins should be sought specifically.
- The orifice between the pulmonary venous chamber and the true left atrium should be measured, and obstruction at this level should be confirmed using color flow and pulsed wave Doppler.
- The presence and degree of *pulmonary hypertension* must be assessed.
- Additional sources of left-sided obstruction should be ruled out

POSTOP:

- Surgical repair of cortriatriatum consists of enlarging the anastomosis between the posterior (pulmonary venous) chamber and the left atrium.
- Postoperative echocardiograms should assess for residual anastomotic site obstruction as well as for individual pulmonary venous obstruction that may have been unmasked following relief of distal obstruction.
- Resolution of pulmonary hypertension is a good indicator of the success of surgery.

Supramitral Ring

- This anomaly is characterized by a membrane at the level of the mitral anulus, which can result in obstruction to left ventricular inflow.
- This is rarely an isolated anomaly; it occurs most frequently in association with other forms of left heart obstruction, including mitral stenosis, aortic outflow obstruction and coarctation of the aorta.
- The membrane may be thin and delicate, representing a subtle finding on transthoracic echocardiography.
- The hemodynamic impact of this lesion is similar to that of mitral stenosis.

ECHO TIP:

- Best shown from parasternal long axis and apical views.
- Should be suspected when prograde transmitral flow acceleration is seen proximal to the mitral valve tips (typically, at the level of the anulus).
- Supramitral ring is related intimately to the mitral valve, being located at or immediately below the level of the mitral anulus. It is oriented anterior and inferior to the left atrial appendage and the foramen ovale.
- The ring may cause tethering of mitral valve leaflets leading to decreased excursion, mimicking mitral stenosis.

POSTOP:

- Surgery for supramitral ring consists of excision of the membrane and repair of any associated defects of the mitral valve.
- Postoperative echocardiograms should assess for residual or recurrent supravalvar obstruction and also for any new or increased mitral regurgitation.
- Resolution of left atrial enlargement and pulmonary hypertension suggest an adequate surgical repair.

Chapter 12

Aortic Valve and Outflow Tract

Aortic Valve & Outflow Tract

- Each of the malformations involving the aortic valve and left ventricular outflow tract may occur either in isolation or in combination with other malformations.
- The importance of these defects lies in their potential to affect left ventricular performance and cardiac output adversely by causing either pressure or volume overload.

Valvar AS
Subaortic Stenosis
Supravalvar AS
Aortic Insufficiency

Valvar Aortic Stenosis

- This lesion is characterized by thickened and/or fused valvar cusps and underdeveloped commissures, leading to stenosis due to inadequate opening of the valve.
- The valve anulus itself may be hypoplastic; multiple levels of left heart obsuucuon may occur in series.
- Stenotic aortic valves are described by the number of cusps (eg., bicuspid or unicuspid) or by the number of commissures (eg., bicommissural or unicommissural).
- In moderate to severe cases, left ventricular hypertrophy is seen.
- Infants with critical valvar aortic stenosis may show left ventricular systolic dysfunction and endocardial fibroelastosis.
- These infants may be dependent on right-to-left flow through the ductus arteriosus for systemic perfusion.

ECHO TIP:

- Valve anulus is best shown from parasternal long axis view.

The morphology of the aortic valve cusps and commissures is best shown from parasternal short axis view with slow playback, examining the systolic opening of the valve cusps.
Transvalvar gradients are best obtained from suprasternal, high right parasternal and apical views.
In all cases, the valve anulus, peak and mean gradient must be measured, with careful attention to the peak velocity proximal to the valve (VI)
Left ventricular systolic and diastolic function and the degree of left ventricular hypertrophy must be assessed.
Multilevel left heart obstructions must be ruled out.
The presence and severity of aortic valve insufficiency must be noted. If aortic insufficiency is present, the location and width of the regurgitant jet should be assessed from parasternal short sxis view.
The location of post-stenotic dilation of the ascending aorta is usually diametrically opposite to the origin of the jet through the stenotic aortic valve.



In short axis view, only two leaflets are shown opening along a single line of closure.



Though flexible, long axis view of a bicuspid aortic valve shows "doming" during systolic opening.

Membranous Sub-aortic Stenosis

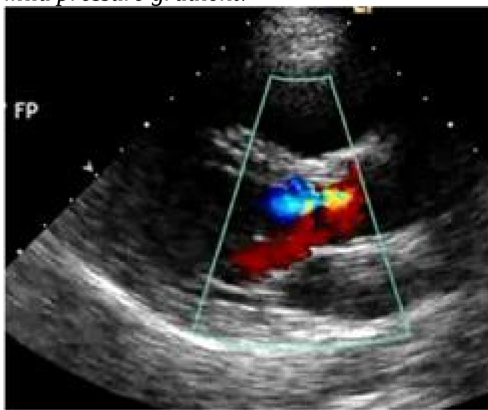
- Subvalvular stenosis occurs in about 10% of aortic stenosis, is more common in males and can be caused by either a discrete membrane or a diffuse fibrous ring in the LVOT ('tunnel).
- Degrees of genetic predisposition are unclear but a pedigree of multiple family members with discrete subaortic membrane has been reported. Varying severities of obstruction occur. The obstructions can create turbulence thickening the aortic valve leaflets and predisposing to endocarditis.

Echo is diagnostically reliable, but careful technique is required since the membrane is often thin. Color doppler will show turbulence.

Transaortic resection of the subaortic membrane is associated with a high incidence of recurrence requiring reoperation (16%). It is reasonable to consider alternative therapies to prevent recurrence in selected cases. Non-surgically, an Inoue balloon catheter has been used retrograde to dilate the membrane, though rupture of a papillary muscle was reported.



Long axis view of the left ventricular outflow tract shows a discrete membrane which creates systolic turbulence and a mild pressure gradient.



Long axis view color doppler of the left ventricular outflow tract shows systolic turbulence and a mild pressure gradient at the subaortic membrane.

Chapter 13

The Aortic Arch

The Aortic Arch

- Normal development of the aortic arch entails the formation of six pairs of primitive aortic arches, one for each branchial cleft.
- These different arches are not all present at one time and various segments regress, with eventual assumption of the appearance of the *normal left aortic arch*.
- Abnormalities of aortic arch continuity, sidedness and
- branching pattern occur with abnormal patterns of formation and regression of the six pairs of aortic arches.

Infantile Coarctation
Discrete Coarctation
Right Aortic Arch
Vascular Ring
Interrupted Aortic arch

Infantile Coarctation

- This lesion is postulated to be due to abnormal development of the left fourth and sixth aortic arches. In addition, contractile tissue from the ductus arteriosus frequently extends into the adjoining portions of the descending thoracic aorta.
- Constriction at the level of ductal insertion can lead to localized coarctation.
- It has also been pointed out that since the lower body is perfused via right-to-left shunting at the level of the ductus, the aortic isthmus in the fetus receives only a smaII fraction of cardiac output. Any lesion that further decreases prograde flow across the isthmus may adversely affect isthmic growth.
- *Thus, infantile coarctation is frequently seen in the setting of a large ventricular septal defect with associated hypoplasia of the aortic outflow tract and anulus.*
- Infantile coarctation is characterized by aortic arch hypoplasia that may include the transverse, proximal and distal arch. The aortic isthmus is frequently long and narrow.
- Localized coarctation is seen as a posterior shelf in proximity to the insertion of the ductus arteriosus.
- Echocardiography is usually adequate for comprehensive diagnosis of infantile coarctation.

Echo Tips:

- Ø Best shown from suprasternal short and long axis views.
- If infantile coarctation is diagnosed, the care providers must be informed promptly to enable institution of prostaglandin El infusion (to maintain ductalpatency) if this has not already been done.
- *The degree to which systemic perfusion is dependent on the ductus arteriosus is assessed by determininig the direction of ductal flow*; thus, right-to-left ductal flow suggests a ductus-dependent lesion.
- Patients with coarctation may have multi-level left heart obstruction and left ventricular hypoplasia which may make biventricular repair difficult or impossible.

- Other sources of aortic outflow obstruction, including supramitral ring, mitral stenosis, subaortic stenosis and valvar aortic stenosis, should be ruled out.
- Measurements of left ventricular volume, left ventricular outflow tract and aortic anulus diameter, and the extent (length and diameter) of arch hypoplasia must be recorded; these can be used in algorithms to determine the feasibility of biventricular repair.
- Ventricular septal defects should be sought using color flow Doppler sweeps of the ventricular septum with a low Nyquist limit, since the pressure gradient across the VSD maybe very small.



Supra-sternal view shows the narrowing of the aorta just proximal to the left subclavian.



Long axis view of the left ventricle and proximal aorta shows the relatively small LV cavity and proximal aorta.

Discrete Coarctation

- This lesion is postulated to be due to abnormal development of the left fourth and sixth aortic arches. In addition, contractile tissue from the ductus arteriosus frequently extends into the adjoining portions of the descending thoracic aorta.
- Constriction at the level of ductal insertion can lead to localized coarctation. Localized coarctation is usually seen as a posterior shelf in proximity to the aortic insertion of the ductus arteriosus; it may also occur in the distal thoracic aorta.
- Echocardiography usually provides comprehensive diagnosis of localized coarctation in infants and young, children. However, in older children, additional imaging modalities such as MRI studies or angiography may be needed,particularly in the unusual situation of distal or thoracoabdominal coarctation.

Echo Tips:

- Best show n from suprasternal short and long axis views.
- Associated defects including bicuspid aortic valve and patent ductus arteriosus must he sought specifically.
- Measurements of aortic arch diameter should be made at several points including proximal arch, distal arch, isthmus, the area of coarctation and the descending aorta distal to the coarctation. The length of the coarctation segment should also be measured.

- These measurements help decide on the feasibility of balloon angioplasty versus surgical repair.
- The presence of diastolic runoff (diastolic persistence of pressure gradient between the segments of the aorta that are proximal and distal to the coarctation) in the absence of a patent ductus arteriosus strongly suggests hemodynamicalIy significant coarctation.

Coarctation – Postop:

- Intervention for coarctation consists of establishing an unobstructed communication from the aortic arch to the descending thoracic aorta.
- This maybe done surgically, with end-to-end anastomosis or left subclavian artery flap repair, or via transcatheter balloon angioplasty.
- Post-intervention echocardiograms should assess for residual or recurrent coarctation.
- Intimal tears, dissecting and true aneurysms at the site of repair should be sought with high frequency transducers.
- Proximal obstruction maybe unmasked by relief of coarctation; therefore, obstruction to prograde flow at the level of the mitral valve, left ventricular outflow tract and aortic valve should all be ruled out.
- Serial echocardiography is important in this lesion.

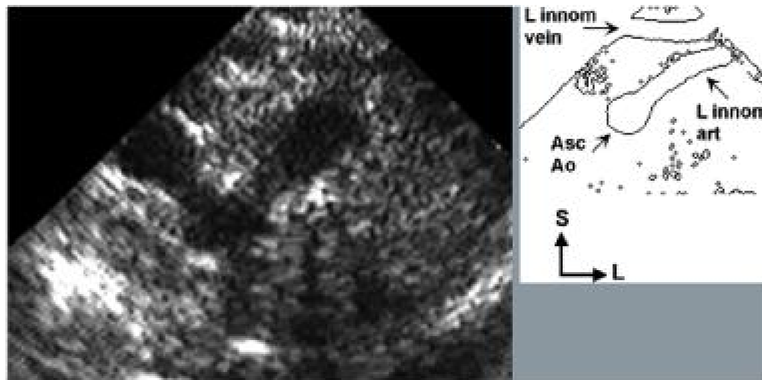
Right Aortic Arch

- Left and right aortic arch refer to which bronchus is crossed by the arch, not to which side of the midline the aortic root ascends.
- Practically, the sidedness of the aortic arch is usually determined indirectly with echocardiography or angiography by the branching pattern of the brachiocephalic vessels. As a rule, the first arch vessel contains the carotid artery opposite the side of the arch.
- A right aortic arch is defined by the course of the aortic arch, which passes to the right of the trachea over the right mainstem bronchus.
- The specific fetal pattern of primitive aortic arch formation and resorption determines the branching pattern of the head and neck vessels. Thus, a right aortic arch may be associated with minor image branching; the braches would then be a left innominate artry, right common carotid artry and right subclavian artery.
- It is important to determine not only to which side of the trachea the aorta passes, but also the branching pattern of the head and neck vessels; this should be achievable by echocardiography in all cases.

Echo Tips:

- Best shown from suprasternal short axis sweep.
- The 'sidedness' of the aortic arch is defined not by its branching pattern but by the side of the trachea that the aorta arches over. Thus, in a patient with a right aortic arch, scanning in suprasternal short axis view and sweeping from anterior to posterior, the lumen of the aorta is seen in cross-section coursing rightwards.
- Once a right aortic arch has been identified, the plane of the suprasternal long axis view should be rotated counter-clockwise to visualize the entire descending aorta.
- Each of the head and neck arteries should be identified from its origin and tracked peripherally. If the first branch off the aorta dose not bifurcate, then it should be traced to identify whether it is a carotid or a subclavian artery. If it is a carotid artery, then the subclavian artery on that side may be arising from the descending aorta,coursing behind the esophagus to reach the ipsilateral arm. This aberrant subclavian artery should be sought diligently.

- If there is an associated patent ductus arteriosus, the origin and insertion of the ductus should be defined.



This suprasternal sax sweep follows the lumen of the ascending aorta and aortic arch posteriorly the aorta arches to the patient right the first branch of the aortic arch goes to the patient left and it bifurcate at the end of the sweep.

Vascular Rings

- *Vascular rings are due to abnormal persistnce or involution of various components of the developing embryonic aortic arches*.
- A right aortic arch with an aberrant retroesophageal left subclavian artery and a double aortic arch comprise the two commonest forms of vascular rings.
- With a *double aortic arch*, both aortic arches are present and are variably patent; they fuse posteriorly to form the descending aorta. In the usual pattern, the right arch is the larger of the two, and is anterior and superior to the smaller left arch .Usually, the two arches give rise to mirror-image branches. *The arches completely surround the trachea*, *forming a complete vascular ring*; the specifics of branching pattern and arch patency determine the clinical manifestations of the vascular ring in individual cases.
- With a right aortic arch, the left subclavian artery may arise anomalously from the descending thoracic aorta, either directly or via a large vessel known as Kommerell's diverticulum. In the latter situation, the diverticulum gives rise to a left-sided patent ductus arteriosus and then tapers to normal size,continuing as the left subclavian artery. In this situation, the trachea is partially surrounded by vascular structures - the pulmonary arterial confluence anteriorly, the left sided ductus to the left, Kommerell's diverticulum posterioly, and the aortic arch on the right. These structures comprise a vascular ring.

Echo Tips:

- Best shown from suprasternal views.
- This defect illustrates the need for determining arch 'sidedness' with a slow anterio/inferior to posterio/superior sweep in *suprasternal short axis view*. This should be followed by a slow left-to-right sweep in *suprasternal long axis* view.
- In suprasternal short axis view, the aortic lumen would be seen to divide into two lumens which continue posteriorly after giving rise to the head and neck vessels. Suprasternal long axis view shows both arches coursing posteriorly; the point where the two arches meet posteriorly may be difficult to demonstrate by echocandiography, and may need additional imaging modalities such as MRI studies.
- The pattern of head and neck vessel branching must be determined by tracking these vessels from their origin.
- The orientation and relative sizes of the two arches must be identified, since the smaller of the two arches is usually divided at surgery.

Vascular Rings – Postop:

- Surgery for double aortic arch consists of division of the smaller (usually the left) arch posteriorly, where the arches meet to form the descending aorta.
- Any patent ductus arteriosus or ligamentum that is identified is also divided.
- Postoperative echocardiogram should assess for any residual vascular ring; this would be very difficult to determine echocardiographically, and other imaging modalities such as MRI are probably indicated. Residual flow through a patent ductus arteriosus should also be sought.

Interrupted Aortic Arch

- In this anomaly, a *segment of the aortic arch is completely absent* .
- This is distinct from severe coarctation of the aorta, where the lumen of the aorta may be occluded but the continuity of the arch structure is maintained. This defect is due to involution of one or more segments that are essential for maintaining normal arch continuity.
- Interrupted aortic arch classification is based on the location of the interruption.
  - *Type A interruption* exists when the interruption is distal to the origin of the subclavian artery that is ipsilateral to the second carotid artery. This type maybe associated with an intact ventricular septum and an aortopulmonory window; this pattern of interruption is also frequently seen with transposition of the great aretries.
  - *Type B interruption* exists when the interruption is between the second carotid and ipsilateral subclavian artery. This type is frequently associated with *DIGeorge syndrome*.
  - *Type C interruption* is a rare type, with interruption between the two carotid arteries.
- Anomalous arch branching patterns are frequently associated with interrupted aortic arch.
- Any form of interrupted aortic arch may be associated with a malalignment type ventricular septal defect .
- The conal septum may be hypoplastic and is malaligned posteriorly and inferiorly into the left ventricular outflow tract.
- Patency of the ductus arteriosus is critical for maintaining perfusion to the lower body in this condition.

ECHO TIP:

- Best shown from suprasternal views.
- If interrupted aortic arch is diagnosed, the care providers must be informed promptly to enable institution of prostaglandin El infusion (to maintain ductal patency) if this has not already been done.
- *The typical superiorly directed termination of the proximal arch differentiates interrupted aortic arch from coarctation of the aorta*.
- The location and length of interruption must be defined.
- A hypoplastic, posteriorly malaligned conal septum, the resulting malalignment type ventricular septal defect, and *size disparity between the dilated pulmonary artery and the small ascending aorta should suggest the diagnosis of interrupted aortic arch*.
- The pattern of arch branching is important for surgical planning. Besides, certain patterns of arch branching lead to less prograde flow across the left ventricular outflow tract (e.g., typeB interruption with anomalous origin of the right subclavian artery from the descending aorta, leaving only the two carotid arteries arising from the ascending aorta) and have been associated with more severe subaortic stenosis from conal septal deviation. Therefore, the pattern of arch branching must be determined in each case.

- Additional aortopulmonary communications, such as patent ductus arteriosus, aortopulmonary window and ventricular septal defect must be sought.

POSTOP:

- Surgery consists of mobilizing the proximal and distal segments of the arch and direct anastomosis of the distal segment to the undersurface of the proximal segment .
- Additional defects, such as patent, ductus arteriosus, ventricular septal defects and aortopuimonary window are closed. Subaortic stenosis due to conal septal deviation may also be resected.
- Postoperative echocardiograms should assess for recurrent or residual obstruction at the site of the arch anastomosis. The adequacy of surgery is limited by the size of the aortic lumen beyond the interruption; thus, the lesion may recur with linear growth of the patient. Therefore, serial echocardiography is important in this lesion.
- Subaortic stenosis is known to recur and worsen in this condition even after adequate surgical repair. The presence and severity of subaortic stenosis must be determined by serial studies.
- If subaortic obstruction is severe and cannot be addressed surgically, the Norwood operation may beperformed, staging the patient for a single-ventricle type of repair. For details on the Norwood operation and staged repair of single ventricle physiology, please go to Chapter 31.

Chapter 14

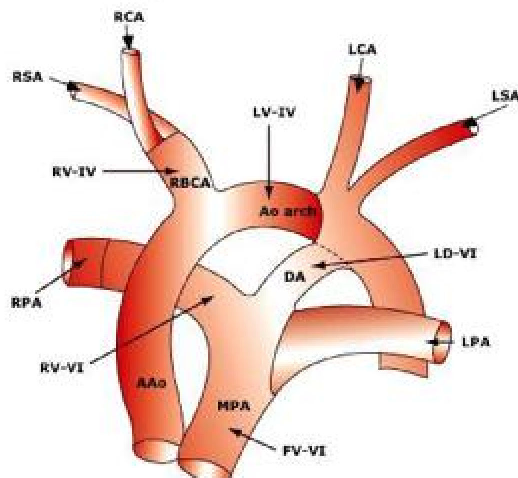
Aortopulmonary Communications

- Communications between the systemic and pulmonary arterial beds maybe due to *persistence of embryonic connections* which should have undergone resorption normally (e.g., patent ductus arteriosus).
- Failure of normal separation of the two outflow tracts also leads to an abnormal communication between the systemic and pulmonary circulations (e.g., aortopulmonary window).
- Finally, abnormal connections may develop between the aorta and the pulmonary parernchyma (e.g., aortopulmonary collaterals).
- These defects lead to a net left-to-right shunt, imposing a volume load on the left ventricle.

PDA
Aortopulmonary Window
Aortopulmonary Collateral

Patent Ductus Arteriosus

- The ductus arteriosus develops from the sixth left aortic arc hand *connects the main pulmonary artery with the descending aorta, distal to the origin of the left subclavian artery*.
- The ductus is normally patent in utero, and usually functionally closes by constriction shortly after birth. Anatomic closure generally occurs by 6-8 weeks of age.
- Patency of the ductus is particularly common in premature neonates. This may occur as an isolated malformation or in association with other cardiac pathology.
- The ductus may also be congenitaly absent as is commonly seen with absent pulmonary valve syndrome or with truncus arteriosus.



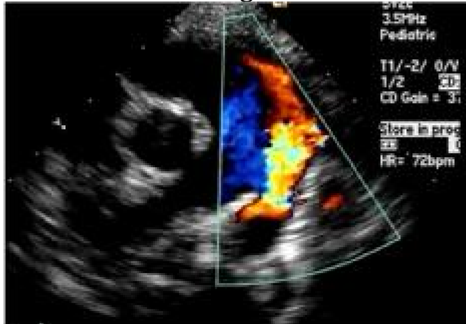
Origin of great vessels and ductus arteriosus

Shown is the morphologic derivation of proximal pulmonary and aortic vessels at birth. The main pulmonary artery (MPA) comes from the fused ventral VIth arterial arch (FV-VI) while the right (RPA) and left pulmonary arteries (LPA) come from the right (RV-VI) and left ventral VIth arterial arches (LV-VI). The ascending aorta (AAo) and right brachiocephalic artery (RBCA)are derived from the right ventral IVth arterial arch (RV-IV), while the aortic arch (Ao arch) comes from the left

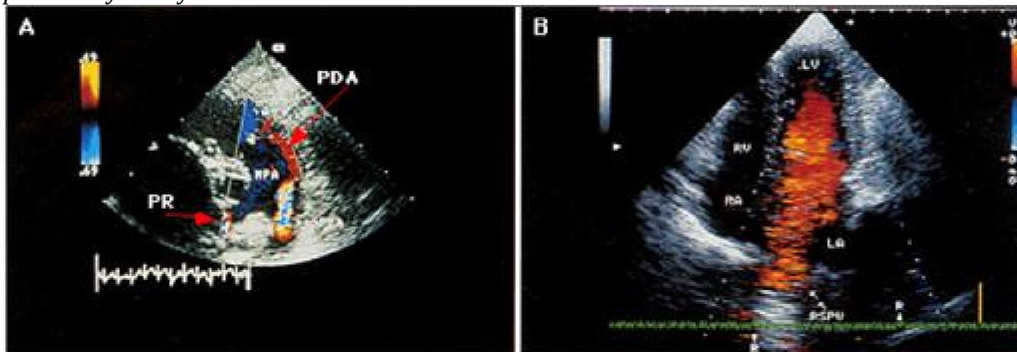
ventral IVth arterial arch (LV-IV). The ductus arteriosus (DA) is derived from the left dorsal VIth arch (LD-VI). LSA: left subclavian artery; LCA: left carotid artery; RSA: right subclavian artery; RCA: right carotid artery.

Echo tip:

- Best seen from high left parasternal (the so-called ductal view), parasternal short axis and suprasternal views.
- A small shunt across the ductus is best shown with color flow Doppler.
- The ductus maybe long and tortuous, making it difficult to image along its entire length.
- Echocardiography should establish the size of the ductus at the aortic and pulmonary ends as well as the length of the ductus and its narrowest diameter.
- Large left-to-right flow across a patent duetus may lead to enlargement, of the left-sided heart chambers.
- Flow patterns in the ductus as assessed by Doppler provide information regarding the direction and velocity of flow, thus enabling measurement of pulmonary artery pressure.
- Aortic arch -sidedness must be established. A right aortic arch maybe associated with either a right-sided ductus leading from the right pulmonary artery to the descending aorta distal to the right subclavian artery, or a left-sided ductus leading from the left pulmonary artery to the proximal left subclavian artery. Aortic arch abnormalities in association with patent ductus or ligamentum arteriosum form the substrate for vascular rings.



Short axis color doppler view at aortic valve level shows flow into the main pulmonary artery from the mouth of the left pulmonary artery.



Color flow Doppler in a patent ductus arteriosus The systolic image of the main pulmonary artery (MPA) shows shunt -'low across a patent ductus arteriosus (PDA) (panel A). The ductal retrograde flow is seen as a bright mosaic jet superimposed on the normal blue flow in the MPA. Note that color away from the transducer is coded blue and flow towards is red. There is also a very small color jet of pulmonary regurgitation (PR) that is in the center of the pulmonary valve. Color flow image in the four chamber apical viev show early diastolic inflow to the left side of the heart due to the large left-to-right shunt (panel B). RV, right ventricle;RA, right atrium; LV, left ventricle; LA, left atrium; RSPV, right superior pulmonary vein.

Postop:

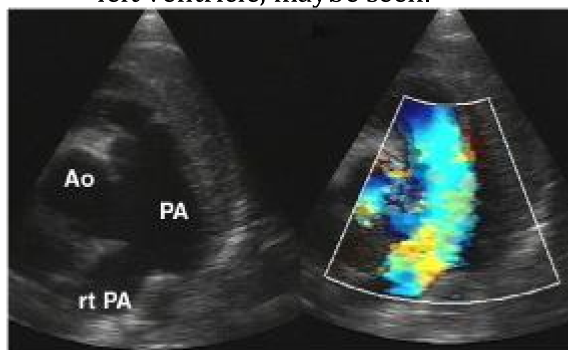
- Treatment of patent ductus arteriosus consists of elimination of ductal patency via surgical ligation/ division or transcatheter coil occlusion.
- Echocardiograms obtained following these procedures should assess for residual shunting as well as for distortion or stenosis of the left pulmonary artery; accidental ligation of the left pulmonary artery has been reported.
- Postoperative echocardiograms should also assess for left ventricular and left atrial size.

Aortopulmonary Window

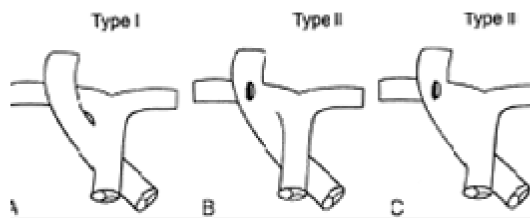
- This lesion is due to a partial defect of the aortopulmonary septum, leading to a communication between the *ascending aorta and the main pulmonary artery*.
- *Aorto-pulmonary Window*, also known as aorto-pulmonary septal defect, is very rare, (0.2 to 0.6 % of all CHD) and is a discrete communication of variable size between aorta and pulmonary artery.
- It is classified according to Mori et. al. by location of the defect into three types:
    - Type 1, the most frequent form, is a communication midway between the semilunar valves and the pulmonary bifurcation.
    - Type 2 is more distal with its distal border is formed by the pulmonary bifurcation; it is commonly associated with an aortic origin of the right pulmonary artery.
    - Type 3 is very rare, consisting of a large and confluent defect involving the entire septum.
- The defect may be seen in isolation or in association with other cardiac pathology such as interrupted aortic arch type A with an intact ventricular septum, tetralogy of Fallot or d-transposition of the great arteries.

ECHO TIP:

- Best seen from parasternal short axis as well as subcostal sweeps at the cardiac base.
- *There maybe a normal area of signal loss in the aortopulmonary septum which may masquerade as an aortopulmonary window. This can be assessed by a parasternal short axis sweep at the base of the heart addressing the area of contact between the ascending aorta and the main pulmonary artery.*
- Associated cardiac defects should be sought.
- The secondary effects of a large left-to-right shunt, including dilation of the left atrium and left ventricle, maybe seen.



Short axis echo labelled on the left shows the aorto-pulmonary communication. The flow from aorta to pulmonary artery is evident in the color doppler imaging shown on the right.



Classification of aortopulmonary window. A: Type I, proximal defect, midway between the semilunar valves and pulmonary bifurcation. B: Type II: distal defect, with posterior border absent and aortic origin of right pulmonary artery. C: Type III, total defect, incorporating defects present in both types I and II.

Postop:

- Surgical repair consists of closure of the defect. In roost instances, the large size of the defect necessitates transaortic approach withpatch closure of the window under cardiopulmonary bypass.
- The unusual small defect maybe amenable to ligation and division without bypass.
- Postoperative echocardiograms should assess for any residual aortopulmonary shunt as well as for distortion or stenosis of the ascending aorta and the main pulmonary artery.

Aortopulmonary Collaterals

- This term refers to extraneous sources of pulmonary blood flow from either the brachiocephalic vessels or the descending aorta.
- Collaterals are frequently seen in situations where progradeflow into the true pulmonary arteries is decreased.
- *The prototype lesion most commonly associated with aortopulmonary collaterals is tetralogy of Fallot with pulmonary atresia.*

ECHO TIP:

- The ability of echocardiography to reliably identify the number, size and distribution of aortopulmonary collaterals is limited.
- A combination of suprasternal and subcostal imaging is useful in detecting collaterals.
- A high index of suspicion for collaterals should be maintained inpatients with malformations such as tetralogy of Fallot with pulmonary atresia.
- Additional forms of imaging such as MRI and angiography *are* more useful than echocardiography.

Postop

- Intervention for aortopulmonary collaterals depends on the distribution of pulmonary blood flow provided by the collateral vessel, and any communication of the collateral bed with the true pulmonary arterial bed.
- Thus, if the true pulmonary artery supplies the same area of lung that is supplied by a collateral vessel, then the collateral maybe occluded to improve the volume of prograde flow, thus stimulating growth of the true pulmonary arteries. Occlusion may be performed by transcatheter coil techniques or by surgical ligation.
- If the pulmonary arteries and collaterals supply separate vascular beds, the repair entails unifocalization of the pulmonary circulation by anastomosing the collateral to the branch pulmonary artery and ligating its aortic connection.
- The role of echocardiography with these complex unifocalization operations is limited to assessment of true pulmonary arterial size proximal to the hilum of the lungs.
- Collateral flow cannot be reliably assessed echocardiographically, and the intraparenchymal details of pulmonary arterial branching and flows are beyond the realm of current transducer technology.

Chapter 15

Hypoplastic Left Heart Syndrome

Hypoplastic Left Heart Syndrome

- This term defines a spectrum of malformations which share underdevelopment of the left heart, including the left atrium mitral valve, left ventricle, aortic valve and aortic arch.
- Circulation depends on the *patency of the ductus arteriosus*, utilizing the right ventricle as the combined systemic and pulmonary ventricle.

Left Atrium, Mitral Valve
Left Ventricle, Aortic Valve
Aortic Arch
Atrial Septum
Right Heart

Left Atrium and Mitral Valve

- The left atrium may be hypoplastic and/or thick-walled, or, less commonly, dilated and thin-walled.
- The mitral valve may be stenotic, hypoplastic or atretic.
- The entire mitral valve apparatus may be abnormally formed, with anular hypoplasia.
- Deficiency of the chordae leading to direct attachment of the mitral valve leaflets to the papillary muscles, obliteration of interchordal spaces, and closely spaced papillary muscles or a single papillary muscle (parachute mitral valve).

ECHO TIP:

- Ø Best show from parasternal long axis and apical views.
- All components of the mitral valve must be assessed systematically.
- The diameter of the mitral anulus, presence and adequacy of chordae tendineae and interchordal spaces, number and spacing of mitral valve papillary muscles must he evaluated.
- The pattern of opening of the mitral valve is best shown in a *apex-to-base sweep in parasternal short axis*.
- The presence of prograde flow across the mitral valve, and the presence and severity of mitral regurgitation must be noted.

Postop

- Surgical options for patients with hypoplastic left heart syndrome consist of staged repair (Norwood series) which culminates in a single (right to systemic) ventricle arrangement . Please go to Chapter 31 for details), or cardiac transplantation with extended reconstruction of the aortic arch (please go to Chapter 30 for details).

Left Ventricle and Aortic Valve

- The left ventricle is almost always hypoplastic; it maybe completely absent. Absence of left ventricular hypoplasia is usually due to an associated ventricular septal defect.

- Left ventricular endocardial fibroelastosis may be seen, particularly if there s prograde flow through the mitral valve in the setting of aortic atresia.
- Atresia of the aortic valve may be due to complete absence of the valve, which is represented by a membrane or shelf, or due to diminutive and imperforate valve structure. When the aortic valve is patent, the leaflets are usually thick and most frequently obstructive.

ECHO TIP:

- Best shown from parasternal long axis and apical views.
- The left ventricle may be diminutive both in absolute size and in comparison to the dilated hypertrophic right ventricle.
- The search for the left ventricle *should be focused around the atrioventricular groove*, since the hypoplastic left ventricle may not develop upto the cardiac apex.
- The presence of left ventricular endocardial fibroelastosis, *seen as echo dense endocardium*, *must be noted*.
- Left ventricular size and systolic function should be assessed. If the mitral valve is atretic and the left ventricle is not correspondingly hypoplastic, additional sources of left ventricular filling such as ventricular septal defects should be sought.

The Aortic Arch and Ductus Arteriosus

- Hypoplasia of the ascending aorta and aortic arch, with or without discrete coarctation of the aorta, is seen in up to 80 % of patients.
- In the classical form of hypoplastic left heart syndrome with mitral and aortic atresia, the ascending aorta is miniscule and serves only as a main coronary artery, with retrograde filling via the ductus arteriosus.
- Patency of the ductus arteriosus allows the right ventricle to provide systemic output; a restrictive ductus would compromise cardiac output.

ECHO TIP:

- Bestshown from suprasternal and high parasternal short axis views. The diameter of the ascending aorta and the proximal and distal aortic arch must be measured.
- Retrograde filling of the aorta from the ductus should be noted.
- The presence, location and severity of aortic coarctation should be defined. *Two-dimensional imaging and color flow Doppler are more helpful* in identifying coarctation than are pulsed or continuous wave Doppler patterns *in the setting of a large patent ductus arteriosus*.

The Atrial Septum

- The morphology of the atrial septum is an important determinant of the clinical manifestations and management outcomes in patients with hypoplastic left heart syndrome.
- Variations of atrial septal morphology have been described including a large secundum atrial septal defect, an intact atrial septum, an aneurysm of septum primum, and leftward malalignment of septumprimum which attaches to the left atrial wall directly, to the left of septum secundum.
- In the presence of an intact atrial septum, pulmonary venous return to the right heart may occur via anomalous pulmonary venous drainage (e.g., Scimitar syndrome, draining to the right atrium), or through the persistence of primitive venous channels between the left atrium and the systemic veins (eg., the levoatriocardinalvein), or an unroofed coronary sinus which allows decompression of the left atrium.

ECHO TIP:

- Ø Best shown from subcostal short and long axis views.
- § Leftward malalignment, and anomalous attachment of septum primum to the left atrial wall should be sought specifically.
- § The location and size of all atrial-Ievel shunts must be documented; this is important for serial assessment as well as for interventions such as balloon atrial septostomy.
- § Small patent foramina ovale are frequently located posterior and superior to their usual location; these should be ruled out using color Doppler.
- § If the atrial septum appears restrictive, dilation of the pulmonary veins should be sought.
- § If the atrial septum is intact, sources of pulmonary venous decompression into the right heart mus t be sought. Each of the pulmonary veins must be tracked. Thus, a subcostal sweep beginning in the transverse abdominal plane sweeping up to the heart will identify *Scimitar syndrome, with anomalous entry of the right-sided pulmonary veins into the right atrium(All the right pulmonary veins or occasionally the veins draining the right middle and right lower lobes enter the IVC either just above or below the diaphragm.)* Echocardiograpic assessment of the coronary sinus septum in the left atrium should help identify a coronary sinus septal defect.
- § Lowering the Nyquist limit for color sweeps from suprasternal imaging should identify levoatriocardinal veins or anomalies of pulmonary venous return.

The Right Heart

- § The right atrium and right ventricle are variably enlarged; this enlargement is more noticeable due to left heart hypoplasia.
- § The right (systemic) ventricle is hypertrophic.
- § Tricuspid valve regurgitation may be present.
- § Right ventricular systolic function may be qualitatively depressed.
- § The pulmonary (neo-aortic) valve may be regurgitant; it is rarely stenotic.
- § Abnormalities of systemic venous drainage may also be present- a fact of importance in planning palliative surgery or cardiac transplantation.

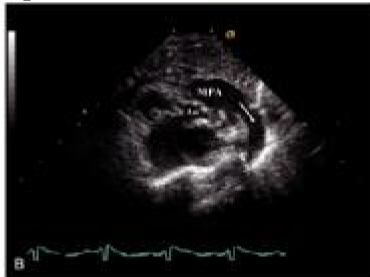
ECHO TIP:

- Ø Best shown from apical and parasternal short axis views.
- § Right ventricular systolic function and Tricuspid regurgitation are difficult to quantify and must be judged qualitatively.
- § Abnormal function (stenosis or regurgitation) of the pulmonary (neo-aortic) valve must be assessed.
- § The surgical implications of systemic venous abnormalities that are identified must be understood; e.g., a persistent left superior vena cava with absence of a left innominate ("bridging') vein may necessitate bilateral Glenn shunts. Thus, systemic venous drainage must be completely defined.

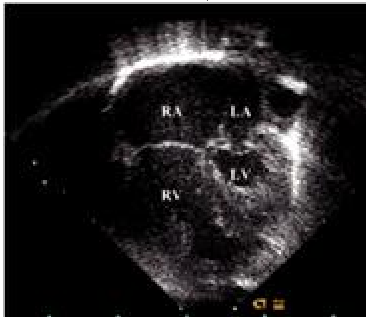


Parasternal long-axis view in a patient with hypoplastic left heart syndrome. The left ventricular chamber is small and muscle bound. The endocardial surface of the left ventricle is echo-bright, consistent with endocardial fibroelastosis (arrow). Ao, aorta; LA, left atrium; RV, right ventricle.

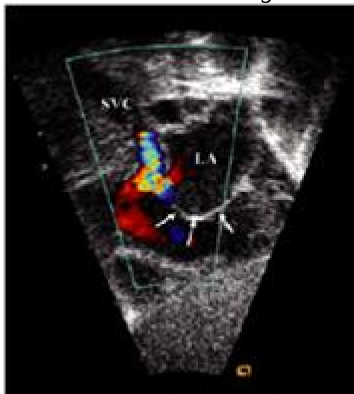




Parasternal short-axis views in a patient with hypoplastic left heart syndrome. In (A) the right ventricle (RV) is enlarged with a smaller, hypertrophied left ventricle (LV). Scanning more superiorly from a parasternal short-axis view (B), the larger main pulmonary artery (MPA) and patent ductus arteriosus (arrow) are seen connecting to the descending aorta. No brachiocephalic vessels are seen arising from the ductal arch, a key finding in differentiating the ductal arch from the true aortic arch. Ao, aorta.



Apical four-chamber view in a patient with hypoplastic left heart syndrome. The left atrium (LA) and left ventricle (LV) are much smaller than the right atrium (RA) and right ventricle (RV). The right ventricle clearly occupies the cardiac apex.



Subcostal sagittal view in a patient with hypoplastic left heart syndrome. The atrial septum is aneurysmal (arrows), bowing into the right atrium. Color Doppler imaging identifies the superiorly positioned atrial shunt with the jet directed into the superior vena cava (SVC). Aliasing of the color Doppler flow signal across the atrial septum is consistent with a restrictive defect and can result in left atrial hypertension. LA, left atrium.

Postop

- Surgical options for patients with hypoplastic left heart syndrome consist of staged repair (the Norwood series) which culminates in a single (right=systemic) ventricle arrangement (please go to Chapter 31 for details), or cardiac transplantation with extended reconstruction of the aortic arch (please go to Chapter 30 for details).

Chapter 16

Truncus Arteriosus

Truncus Arteriosus

- Truncus Arteriosus accounts for 1-4% of all CHD, is more frequent in males and is associated with extra-cardiac anomalies in 20-30 % of cases. It is characterized by a single arterial trunk arising from the heart supplying the systemic, pulmonary and coronary circulation, and results from a defect in cono-truncal septation. This accounts for the almost universally large ventricular septal defect. The truncal valve may have one, or as many as six cusps - but is most commonly tricuspid, quadricuspid or bicuspid. In addition, the truncal valve may be stenotic and is often insufficient, sometimes significantly.
- This defect is due to failure of normal septation of the embryonic truncus arteriosus into the aorta and main pulmonary artery.
- In this malformation, the heart has a single semilunar valve; a single artery originates above this valve and *gives rise to the coronary, pulmonary and systemic arterial circulation*.
- There is *almost always a ventricular septal defect* due to absence of the conal septum; the defect is therefore located in the 'Y' of the septal band.
- The truncal valve usually exhibits dysplasia of the cusps and commissures, with varying numbers of commissures and varying degrees of valvar stenosis and/or regurgitation.
- Aortic arch abnormalities such as a right aortic arch are common. Interrupted aortic arch is seen less frequently.

VSD
Truncal Valve
Aortic Arch Abnormalities
Pulmonary Arteries (Classification)

Ventricular Septal Defect

- A ventricular septal defect is *almost always present* in association with truncus arteriosus.
- The VSD is due to absence of the conal septum; thus, it is located in the *outflow portion* of the heart. It is bordered by the two limbs of the septal band inferiorly, and by the truncal valve itself superiorly.

ECHO TIP:

- Best shown from parasternal and subcostal views.
- Frequently associated with prolapse of truncal valve cusps into the defect, which may partially close the defect and may also cause truncal insufficiency: truncal valve cusp mobility and coaptation should be assessed.
- Defect location, margins and extent should be determined.
- Sizing of the defect should be performed at end-diastole in orthogonal planes using 2-D imaging as well as color.
- Chordae or papillary muscles straddling the defect should be identified.
- Structures that may impede surgical visualization of the entire extent of the defect,such as prominent right ventricular muscle bundles, should be noted.
- The color sweep of the ventricular septum that is routinely performed to rule out additional VSDs should utilize a low color scale (Nyquist limit).

Postop

- The goals of surgery for truncus are to achieve biventricular repair, separating the pulmonary and systemic circulations. Surgery consists of closing the ventricular septal defect to the aorta; the pulmonary arteries are detached from the aorta and connected to tie right ventricle with a homograft conduit. Repair or replacement of the truncal valve is performed if needed.
- Postoperative echocardiograms should assess for residual VSD, right ventricular systolic pressure, conduit obstruction along the entire length of the conduit, and distortion or stenosis of the branch pulmonary arteries. The truncal valve should be examined for regurgitation or stenosis.

Truncal Valve

- The truncal valve most frequently overrides the ventricular septum.
- The valve itself is frequently dysplastic, and may exhibit thickened cusps, hypoplastic commissures leading to stenosis and/or imperfect cusp coaptation leading to regurgitation.

ECHO TIP:

- Ø Best shown from parasternal short axis and parasternal long axis.
- The number of cusps and commissures, the adequacy of commissural opening in systole and cusp coaptation or prolapse in diastole must be evaluated.
- A careful color sweep of the truncal valve in short axis view should assess for the origin of any regurgitant, jets.

Postop

Same as above.

The Aortic Arch

- Patients with truncus arteriosus frequently exhibit abnormalities of the aortic arch, including right aortic arch and interrupted aortic arch type B.
- The ductus arteriosus may be patent; in patients with interrupted aortic arch, it is frequently large and may be mistaken for the true aortic arch.

ECHO TIP:

- Ø Best shown from high parsternal and suprasternal views.
- Identifying an interrupted aortic arch with truncus arteriosus is a diagnostic challenge.
- In order to assess for interrupted aortic arch, the lumen of the ascending aorta should be followed in cross-section in the coronal plane, sweeping posteriorly.
- The true aortic arch is frequently quite small, especially in comparison to the size of die ductal arch.
- *In the suprasternal long axis view, absence of the origin of any brachiocephalic vessels from the ductal arch helps to avoid mistaking the ductal arch for the true aortic arch.*
- Each of the arteries supplying the head and neck must be identified, since there may be associated abnormal arch branching patterns.

Postop

Same as above

Pulmonary Arteries (Classification)

- Collett and Edwards' classification of truncud arteriosus is based on the pattern of origin of the pulmonary arteries from the truncus.

- *In type I*, the main pulmonary artery arises from the truncus and divides into the right and left pulmonary arteries.
- *In type II*, there is no identifiable main pulmonary artery. The branch pulmonary arteries arise from the truncus separately, *but their ostia are close together*.
- *In type III*, the branch pulmonary arteries arise from the *ipsilateral aspects* of the truncus.
- The commonest form of truncus arteriosus encountered clinically is one where the mam pulmonary artery is very short, giving rise to the branch pulmonary arteries almost immediately.

ECHO TIP:

- Best shown from subcostal long axis and parasternal short axis sweeps.
- The pulmonary arteries arise *almost immediately above the truncal valve*, most frequently from the leftward posterior aspect of the truncus.
- The main, right and left pulmonary arteries should each be assessed separately. Stenosis at any site should be identified.
- If the main pulmonary artery is very short, it may be challenging to differentiate type I from type II truncus. Careful sweeps at the base of the heart are indispensable for an accurate diagnosis.

Post op

Same as above



Labelled short axis view at the level of the great vessel shows by the arrow the connection between the aorta (Ao) and pulmonary artery (PA).

Chapter 17

d-Transposition Of The Great Arteries

Atrioventricular Concordance and Ventriculoarterial Discordance or
Simple Transposition of the Great Arteries

d-Transposition Of The Great Arteries

- Transposition of the great arteries is defined as ventriculoarterial discordance, that is, the aorta arises from the morphologic right ventricle and the pulmonary artery arises from the morphologic left ventricle.
- TGA is most commonly seen in the setting of visceroatrial situs solitus and ventricular d-looping.
- The embryologic basis of this conotruncal malformation is the subject of controversy. *One widely accepted theory is that transposition is due to persistence of the subaortic conus and involution of the sobpulmonary couns*. Persistence of the subaortic conus lifts the aortic valve anteriorly, superiorly and rightwards relative to the pulmonary valve, and leads to alignment of the aorta with the right ventricle. Absence of the sobpuhnomry conus leads to mitral-pulmonary continutty and aligimient of the pulmonary artery with the left ventricle.

Great Arteries
VSD
Outflow Tracts

AV Connection	*VA Connection*	*Defect*
Concordant	Concordant	Normal heart
Concordant	Discordant	d-Transposition
Discordant	Discordant	l-Transposition Isolated
Discordant	Concordant	ventricular inversion
		Anatomically corrected malposition

- In the most common form of transposition the morphologic right atrium on the patient's right is connected to the morphologic right ventricle on the patient's right, which in turn is connected to the aorta. On the patient's left the morphologic left atrium is connected to the morphologic left ventricle, which is connected to the pulmonary artery. This *defect is called situs solitus*, *d-loop*, *d-transposition,or simply d-transposition*. The "*d*" in the third term is used to describe the spatial relations of the aortic and pulmonic valves.

- The mirror image of this defect is situs inversus, *l*-loop, *l*-transposition. In more than 80% of cases the aortic valve spatially is located to the right of the pulmonary valve; hence, the use of the "*d*."
- Other spatial relationships are possible. In a small percentage of cases the aortic infundibulum and valve may be to the left of the pulmonary valve. This spatial relationship is frequently found in patients with *d*-transposition and a large ventricular septal defect, and it is often referred to as situs solitus, *d*loop,*l*-transposition. In this nomenclature the "*l*" in the third term refers to the spatial position of the aorta (to the left). In another small percentage of patients the aortic valve may be directly anterior to the pulmonary valve.
- *The echocardiographic diagnosis of transposition of the great arteries is based on demonstrating an abnormal connection of the right ventricle to the aorta*. Although the spatial relationships of the great arteries may provide supportive evidence of the diagnosis, they should never be used as the sole diagnostic criteria.
- The connections between the ventricles and great arteries can be seen in multiple echocardiographic views; *however, the subcostal views are particularly useful for complete segmental analysis of the heart* .

ECHO TIP:

- The connections between the ventricles and great arteries can be seen in multiple echocardiographic views; *however, the subcostal views are particularly useful for complete segmental analysis of the heart* .
- View obtained by tilting the transducer posteriorly to image the inlets of the ventricles. *The eustachian valve can be imaged in the morphologic right atrium (RA) and the pulmonary veins can be seen draining to the morphologic left atrium (LA).*Then plane of sound tilted slightly anteriorly to image the midportion of the heart. The smooth-walled morphologic left ventricle (LV) on the patient's left (d-loop) gives rise to a posterior vessel that bifurcates and is therefore the pulmonary artery (PA).Then plane of sound tilted far anteriorly. The morphologic right ventricle (RV) on the patient's right gives rise to the aorta (AO).
- *Anatomic features include* (1) complete subaortic muscular infundibulum,pulmonary valve-mitral valve fibrous continuity, (3) parallel spatial alignment of the outflow tracts and great arteries, (4) straight muscular septum, and posterior angulation of the posterior great artery.
- *Associated Defects:*

Ventricular Septal Defect.

- One of the objectives of echocardiography is to define the feasibility of biventricular repair, which can be complicated in the presence of a ventricular septal defect. VSD's occur in about a third of patients with {S ,D ,D} T G A.
- Defect location is variable; *malalignment-type defects are common*. Posterior, inferior and rightward deviation of the conal septum can lead to left ventricular (pulmonary) outflow tract obstruction associated with valvar pulmonary stenosis, potentially making these patients ineligible for an arterial switch operation wherein the left ventricular outflow tract would become the neo-aortic outflow tract.
- Anterior, superior and leftward deviation of the conal septum can lead to right ventricular (aortic) outflow tract obstruction. *This can be associated with aortic valvar hypoplasia and coarctation of the aorta, which must be identified preoperatively*.
- Canal-type or inlet VSD may also occur inpatients with TGA. These defects maybe large VSDs may be associated with straddling of the tricuspid valve across the interventricular septum.

Since these attachments may make biventricular repair difficult or impossible, they must be specifically sought in each case.

- Muscular VSD (single or multiple) can also occur in patients with TGA.
- Ventricular septal defects occur in about 33% of patients with *d*transposition.*Most of these defects are in the outlet septum and are associated with an overriding pulmonary artery*. In this situation, anterior displacement of the infundibular septum results in a narrowed right ventricular infundibulum, discontinuity of the infundibular septum and the trabecular septum, and a *malaligned-outlet ventricular septal defect*.
- Although the pulmonary artery overrides the ventricular septum, more than 50% of the pulmonary artery is committed to the left ventricle and there is *pulmonary-mitral continuity*.
- In patients with a *malaligned-outlet ventricular septal defect*, *tricuspid valve abnormalities are frequent*,occurring in 65% of patients in one series.The types of tricuspid valve anomalies that occur are
    - chordal attachments to the infundibular septum or ventricular septal crest,
    - overriding of the tricuspid annulus, straddling tricuspid valve with chordal attachments into the left ventricle,
    - tricuspid valve tissue protruding through the ventricular septal defect and causing subpulmonary obstruction,
    - and cleft anterior leaflet of the tricuspid valve.
- The anterior displacement of the infundibular septum causes subaortic narrowing and produces a long, oblique course from left ventricle to aorta.
- These anatomic features make intraventricular repair extremely difficult and favour repair with an arterial switch procedure, closure of the ventricular septal defect, and resection of subaortic muscle, if necessary.After closure of the ventricular septal defect, the right ventricle always becomes smaller, and right ventricular outflow gradients that were of minor significance preoperatively may become significant.
- The *subaortic narrowing* seen in patients with *d-transposition and a malaligned-outlet ventricular septal defect* may lead to the development of coarctation and interruption of the aorta.
- Other types of outlet ventricular septal defects occur less commonly in patients with *d*-transposition. *Outlet ventricular septal defects can occur with posterior displacement of the infundibular septum*, left ventricular outflow tract narrowing, and posterior malalignment between infundibular septum and trabecular septum.In these cases, muscular subpulmonary obstruction is nearly always present. Because of the posterior deviation of the infundibular septum, a direct route from left ventricle to aorta is present, and patients with this defect are good candidates for repair by way of intraventricular rerouting from left ventricle to aorta. Coarctation of the aorta is not associated with this type of ventricular septal defect.
- Another type of outlet ventricular septal defect that occurs very infrequently in patients with *d*-transposition is a *subarterial (subaortic) ventricular septal defect in which the infundibular septum is hypoplastic or absent*, *but not displaced*. In patients with this defect, the aorta is frequently to the left and anterior.
- *Perimembranousinlet ventricular septal defects* are commonly found in patients with *d*-transposition. This type of defect is also associated with tricuspid valve abnormalities of the type described earlier for a malaligned-outlet defect. In one series, 100% of patients with inlet ventricular septal defect had tricuspid valve abnormalities.
- Other types of defects found with *d*-transposition are isolated muscular defects and perimembranous trabecular defects.

ECHO TIP:

- Ø Best seen from subcostal and apical views.
- § *To identify structures that may impede future VSD closure to the aorta, the VSD must be profiled from views that simultaneously show the aortic valve*. Structures that cross the VSD in these planes may complicate defect closure, and must be defined.
- § To ensure that there are no A-V valve attachments straddling the ventricular septum, sweeps from multiple planes of interrogation at the level of the VSD must be used.
- § To identify potential sources of postoperative outflow obstruction, preoperative echocardiograms must include a mental picture of the orientation of the VSD patch.
- § Additional VSDs should be ruled out with careful color sweeps of the ventrivular septum; a low Nyquist limit maybe needed.
- § Additional levels of intracardiac shunting (at the level of the atrial septum and the ductus) must be sought and their magnitude estimated.
- § The presence of a malaljgnment-type VSD should lead to the search for outflow tract obstruction. Multiple levels of outflow tract obstruction must be ruled out.

Left Ventricular Outflow Tract Obstruction.

- § Fixed ventricular outflow tract obstruction can be due to subvalvar membranes, fibromuscular ridges, atrioventricular valve attachments or semilunar valve stenosis. Inpatients with a VSD, malalignment of the conal septum can lead to outflow tract obstruction.
- § Obstruction to the pulmonary outflow occurs in patients with *d*transposition with or without a ventricular septal defect.
- § With an intact ventricular septum, dynamic subpulmonary obstruction is common, either before or after an intra-atrial baffle procedure. This dynamic obstruction is caused by a prominent systolic bulging of the ventricular septum into the left ventricular outflow tract.
- § On the short-axis views the *left ventricle is thin walled and crescent shaped*.
- § Only minimal pressure gradients are detected by pulsed or continuous wave Doppler techniques.
- § With significant *fixed anatomic obstruction* and an intact ventricular septum, the left ventricular pressure increases and the left ventricle becomes *spherical and thick walled*. The most common forms of fixed pulmonary stenosis in this setting are fibrous subpulmonary diaphragm, fibromuscular ridge, and valvular stenosis (usually a bicuspid pulmonary valve).
- § Subpulmonary stenosis occurs more commonly in association with a ventricular septal defect, and certain types of subpulmonary stenosis are specific for certain types of ventricular septal defect. For example, posterior deviation of the infundibular septum occurs with malaligned-outlet ventricular septal defect, whereas accessory tricuspid leaflet tissue bowing into the left ventricular outflow tract tends to occur with perimembranous-inlet defects.

ECHO TIP:

- Ø Best seen from subcostal and parasternal short and long axis views.
- § Actual obstruction as well as the potential for future (postoperative) obstruction must be assessed.
- § In the presence of a VSD, anatomic definition of the substrate for obstruction is more meaiungful than are Doppler gradients.

- If one level of outflow tract obstruction is found, additional levels of obstruction must be sought.

Great Arterial Relationships

- The position of the aortic valve relative to the pulmonary valve is variable. Anterior and rightwards malposition of the aortic valve is commonest. Five variations are possible:

1) *D-malposition of aortic valve*: Persistence of subaortic conus and absence of (or rudimentary) subpulmonary conus, with the *aortic valve anterior and rightwards of the pulmonary valve.*
2) *Side by side great arteries*: Persistence of subaortic and subpulmonary (bilateral) conus, with the aortic valve usually located to the right of the pulmonary valve in the same anterior-posteriorplane.
3) *Solitus arrangement*: Normal conal anatomy, with persistent subpulmonary conus and absent or rudimentary subaortic conus; the aortic valve is posterior and rightwards of the pulmonary valve. This feads to TGA with posterior aorta, a very rare occurence.
4) *Inverse arrangement*: Usually seen with situs inversus. Persistence of subpulmonary conus and absent or rudimentary subaortic conus; the aortic valve is posterior and leftwards of the pulmonary valve. This is very rare.
5) *L-malposition of the aortic valve*: Persistence of subaortic conus, absent or rudomentary subpulmonary conus. The aortic valve is anterior and leftwards of the pulmonary valve.

ECHO TIP:

- Ø Best shown from subcostal sweeps and high parasternal short axis views.
- Very rarely, transposition can occur with a posterior aorta. Therefore, the anterior-posterior relationships of the semilunar valves should not be used to diagnose transposition.
- Side by side arrangement of the great arteries is more likely to be associated with abnormal coronary artery patterns. 'While we have not discussed coronary artery anatomy in transposition in this presentation, the subject has been discussed comprehensively in several published articles”.

Postop

- The choice of surgical procedures for transposition depends on the associated defects. In the absence of complicating issues such as outflow tract obstruction or either a malalignment type VSD or noncommitted (e.g., canal-type) VSD, the arterial switch operation is currently the procedure of choice.
- Before the arterial switch was perfected, surgery for TGA consisted of an atrial-level switch operation (Senning or Mustard). For details on switch p rocedures undertaken for patients with transposition of the great arteries, please go to Chapter 32 (Switch Procedures).
- Switch operations may not be an option in the presence of outflow tract obstruction. In these situations, the Rastelli operation may be used or modified to achieve biventricular repair. For details on the Rastelli operation, please go to the Postop section in Chapter 17 (Double Outlet Right Ventricle).
- Straddling atrioventricular valves may make biventricular repair difficult or impossible. In that situation, the patjent may need to be staged towards the Fontan operation. For details on the Fontan series of operations, please go to Chapter 31 (Staged Repair - Single Ventricle).



Long axis shows the posterior pulmonary artery and anterior aorta aligned rather than in the usual crossing arrangement.


Short axis shows the posterior pulmonary artery and anterior aorta aligned rather than in the usual crossing arrangement.


Four chamber view shows the large anterior ventricle which can be identified as a morpholgic right ventricle because of its trabeculation and moderator band. Structures from a Mustard repair of the atria can also be noted.

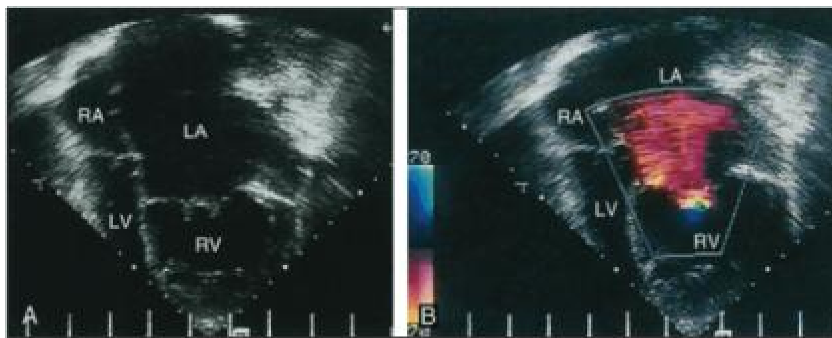
Chapter 18

L-Transposition Of The Great Arteries

Atrioventricular Discordance and Ventriculoarterial Discordance or l-Transposition of the Great Arteries

- This malformation is also known as *'congenitally corrected transposition of the great arteries' or ventricular inversion with l-transposition of the great arteries'*.
- *l*-Transposition of the great arteries is a condition in which there is both atrioventricular and ventriculoarterial discordance; therefore, *in situs solitus* the morphologic right atrium on the patient's right is connected to the morphologic left ventricle on the patient's right, which in turn is connected to the pulmonary artery. On the left the morphologic left atrium is connected to the morphologic right ventricle, which is connected to the aorta. This defect is called *situs solitus*, *l-loop*, *l-transposition*, *or simply l-transposition*. The "*l*" in the third term is used to describe the spatial relations of the aortic and pulmonic valves.
- These hearts exhibit atrioventicular discordance and ventriculoarterial discordance.
- In most cases the *aortic valve is to the left; hence the use of the "l."* Because of the presence of discordant connections at two levels, the circulation is hemodynamically correct (systemic venous blood flows to the pulmonary artery and pulmonary venous blood flows to the aorta); some investigators have called this defect *corrected transposition of the great arteries. The mirror-image situation is situs inversus*, *d-loop*, *d-transposition*.
- As with *d*-transposition, the echocardiographic diagnosis of *l*-transposition is based on demonstrating abnormal connections between the right ventricle and aorta and also between the atria and the ventricles.
- Thus, in a patient with visceroatrial situs solitus, the right atrium opens via the right sided atrioventricular (morphologic mitral) valve into the right sided (morphologic left) ventricle, which connects to the main pulmonary artery via the pulmonary valve which is rightward and posterior of the aortic valve, usually without a subpulmonary conus.
- The left atrium opens via the morphologic tricuspid valve into the morphologic right ventricle, which connects to the aorta via the aortic valve which is supported by a subaortic conus, lifting the aortic valve anteriorly and leftwards of the pulmonary valve.(Normal conal (infundibular)) anatomy is characterized by a *subpulmonary conus* and absence of a subaortic conus)
- Thus, although the internal aligmments are discordant and transposition is present, the circulation is 'corrected'; hence the alternative forms of nomenclature.
- This malformation is due to abnormal leftward (L) looping of the straight heart tube in the embryo, which brings the morphologic left ventricle to lie rightward of the morphologic right ventricle.
- A variety of defects are associated with this malformation Ventricular septal defect is the commonest associated lesion; these defects can vary in location, size and number.
-
- *Eho tip:*
  - In the Subcostal coronal

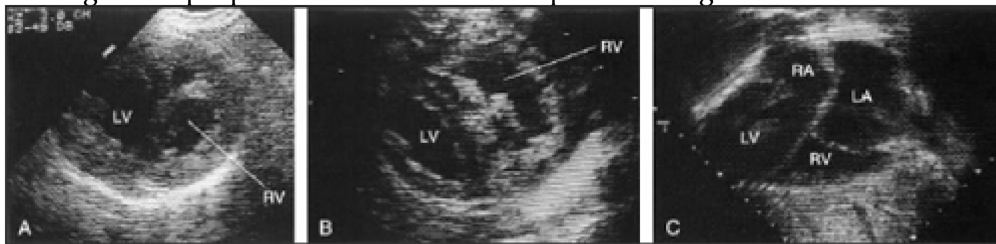
- ü The transducer tilted posteriorly to image the inlets of the heart. The pulmonary veins can be seen draining to the left-sided atrium, indicating that it is the morphologic left atrium (LA) and there is atrial situs solitus.
- ü Then plane of sound then tilted anteriorly. The right-sided ventricle has a smooth septal surface and a shape suggesting that it is the morphologic left ventricle (LV). The LV gives rise to a vessel that bifurcates into two branches and is the pulmonary artery (PA). These findings indicate an l-loop with l-transposition.
- ü Note if there is subvalvular and valvular pulmonary stenosis and the poststenotic dilation of the PA.
- ü If there is an outlet ventricular septal defect.
- ü Then plane of sound tilted far anteriorly. The ventricle on the left side is triangular in shape and has prominent septal-parietal free wall muscle bundles, indicating that it is the morphologic right ventricle (RV). The RV gives rise to a vessel that arches and is the aorta (AO).



A, Apical four-chamber view from a patient with atrial situs solitus, l-loop, and l-transposition of the great arteries.Note that the ventricle on the left has a prominent moderator band and an atrioventricular valve closer to the cardiac apex. These findings indicate that the leftsided ventricle is the morphologic right ventricle (RV) and there is an l-loop. The leftsided tricuspid valve in this patient is much more apically displaced than is normal because of associated Ebstein deformity of the valve. B, Color Doppler examination from the same view shows a tricuspid regurgitation jet (red flow area) with a wide proximal diameter indicating a large regurgitant orifice. Note the massive left atrial (LA) dilation caused by the physiologic mitral regurgitation. LV, left ventricle; RA,right atrium.

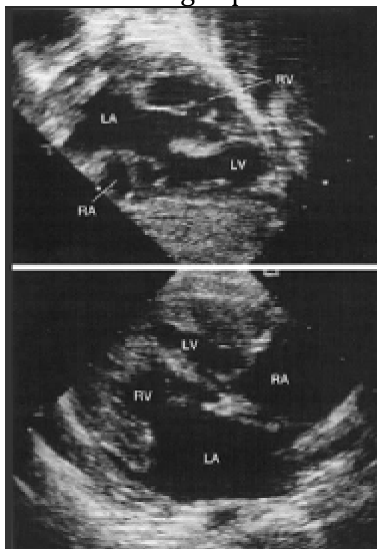
- § The spatial relationships of the great arteries may provide supportive evidence of the diagnosis, but these are never used as the sole diagnostic criteria. *In l-transposition the aortic valve is usually supported by a complete muscular infundibulum and is therefore located more superiorly than the pulmonary valve .*
- § In most cases the muscular infundibulum of the left ventricle is absorbed so that the pulmonary valve is wedged deeply in the heart between the two atrioventricular valves. Direct valvular continuity may exists between the posterior cusp of the pulmonary valve and the anterior leaflet of the mitral valve; however, indirect continuity also exists with the tricuspid valve on the left via the central fibrous body and membranous septum.*With a slight tilting of the plane of sound in the parasternal longaxis view, "continuity" of the pulmonary valve to both atrioventricular valves can be shown.*
- § As in *d*-transposition, the ventricular outflow tracts and great arteries in *l*transposition exit the heart in a parallel fashion rather than wrapped around each another. *Unlike the normal heart or the heart with d-transposition, however, the right ventricle is usually not anterior to the left ventricle in l-transposition.*

- Typically, the ventricles are positioned side by side and the ventricular septum is oriented in a straight line perpendicular to the frontal plane through the thorax .



A, Parasternal short-axis view from a patient with situs solitus, l-loop, and l-transposition of the great arteries with partial apical pivoting. The ventricle on the left has a prominent moderator band and is the morphologic right ventricle (RV). Note that the ventricles are positioned side by side,with the septum oriented perpendicular to the frontal body plane. This arrangement of the ventricular mass and septum is due to lack of complete apical pivoting, which commonly occurs in hearts with atrioventricular discordance. B, Parasternal short-axis view from a patient with situs solitus, l-loop, and l-transposition of the great arteries with complete apical pivoting. The apex has pivoted completely to the side opposite the loop and thus points to the right hemithorax. As a result, the ventricular septum is oriented in a plane the mirror image of normal. C, Subcostal coronal view from a patient with situs solitus, l-loop, and l-transposition of the great arteries with complete apical pivoting. The apex has completely rotated to the side opposite the loop. Note the left atrioventricular valve positioned closer to the cardiac apex, indicating that it is a tricuspid valve. The plane of the ventricular septum is the mirror image of normal.LA, left atrium; LV, left ventricle; RA, right atrium.

- In some cases the ventricles are arranged in a superoinferior fashion, the morphologic right ventricle being superior .



Top, Subcostal coronal view from a patient with situs solitus, l-loop, and l-transposition and superior-inferior arrangement of the ventricles. The left atrium (LA) communicates by way of a straddling and overriding tricuspid valve to a superiorly positioned right ventricle (RV). Note the horizontal position of the ventricular septum and the inferiorly positioned, smooth-walled left ventricle (LV). Bottom, Parasternal view through the inflow tracts of both ventricles.
The two atrioventricular valves are seen in the same view aligned parallel, indicating that this patient does not have crisscross atrioventricular relations. RA, right atrium.

- The unusual spatial relationships of the ventricles, ventricular septum, and great arteries can lead to unusual and often confusing echocardiographic images (especially in the parasternal views).

Associated Defects:

Ventricles & Great Arteries
Ventricular Septal Defect.
Tricuspid Valve
Outflow Tracts

Ventricles and Great Arteries

- Ventricular l-looping is diagnosed on the basis of the left ventricle being to the right of the right ventricle, as discussed in chapter 3 (Segmental Anatomy).
- Thus, the normal mitral-tricuspid offset is reversed, so that the septal hingepoint of the left sided A~V valve is apically displaced(toward the apex) compared to that of the right-sided A-V valve.
- The left-sided A-V valve is septophilic, with chordal attachments to the ventricular septum, and it does not exhibit continuity with the aortic valve.
- The right-sided A-V valve is septophobic with no septal attachments, and it exhibits continuity with the pulmonary valve.
- The left-sided (morphologic right) ventricle has a trabeculated septal surface, while the right-sided (morphologic left) ventricle has a smooth septal surface.
- The plane of the ventricular septum is more sagitally oriented than is normal, and the great arteries are parallel in their course.
- The aortic valve is usually leftward and anterior to the pulmonary valve.

ECHO TIP:

- Best shown from subcostal sweeps.
- The parasternal long axis view is oriented more vertically than normal.
- The parastemal short axis view is oriented more horizontally than normal.
- The ventricular septum is oriented more sagittal than normal.
- The great arteries are parallel rather man spirally twisted, and can be imaged simultaneously in the parasternal long axis view.

Postop

Patients with atrioventricular discordant transposition of the great arteries may need surgery for the associated defects; thus VSD closure,relief of subpuhnonary obstruction and tricuspid valve repair or replacement maybe indicated for individual patients. Postoperative echocardiograms would therefore need to be individualized depending on the specific operation performed.

Ventricular Septal Defect.

- Ventricular septal defect occurs in about 70% of patients with *l*transposition and is *usually perimembranous* in location. In l-transposition,ventricular septal defects are frequently accompanied by other malformations (i.e., perimembranous inlet defects are associated with tricuspid valve straddle; anterior outlet defects are associated with mitral valve straddle).

The Tricuspid Valve

- The left-sided tricuspid valve is frequently thickened and dysplastic; the chordae tendineae may be abnormally short and tethered, with diminution or even obliteration of the interchordal spaces.

- Apical displacement of the septal and/or posterior leaflets of the tricuspid valve may be present. - the so-called Ebsteinoid malformation (of the the left-sided tricuspid valve).
- Tricuspid regurgitation is frequently seen. The tricuspid valve may also straddle across a canal-type (inlet) ventricular septal detect.

ECHO TIP:

- Regurgitation and reversed offset of the A-V valves is best shown from apical views.
- Color flow Doppler interrogation of the tricuspid valve is important for assessing the severity of tricuspid regurgitation.
- A comprehensive assessment of the entire tricuspid valve must be performed, including anulus size, apical displacement, adequacy of leaflet coaptation, length of chordae tendinaeae, presence and adequacy of interchord al spaces, number and spacing of papillary muscles.
- Straddle of the tricuspid valve across a ventricular septal defect must be ruled out.

The Outflow Tracts

- Left ventricular (pulmonary) outflow obstruction is common. This maybe due to atrioventricular (mitral, or tricuspid accessory atrioventricular tissue through a VSD) .
- Absence of the subpulmonary conus pulls the pulmonary valve and outflow tract down between the ventricular *free* wall and the conal septum, leading to dynamic subpulmonary obstruc tion. Outflow tract obstruction can occur at multiple levels, from the subvalvuar outflow tract to the branchpuhnonary arteries. Aortic outflow tract obstruction is rarely seen in this malformation.

ECHO TIP:

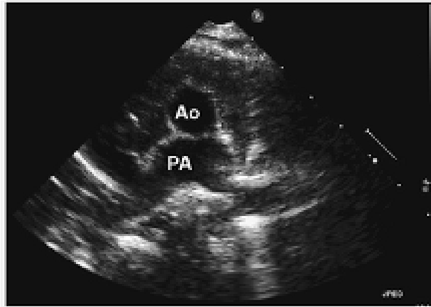
- Best shown from subcostal long axis sweeps and parasternal long axis views.
- The location, mechanisms and severity of outflow tract obstruction must be determined to allow assessment of the feasibility of intervention.
- The absence of a pressure gradient across an outflow tract is meaningless in the presence of a ventricular septal defect; in this setting, two-dimensional imiaging of the outflow tract is very useful.

Postop

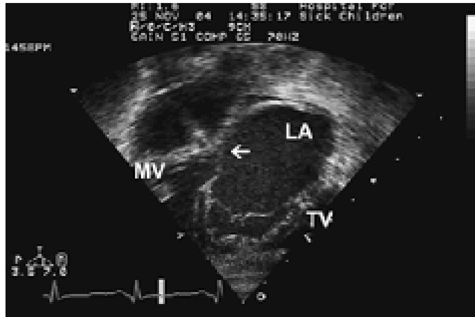
Patients with atrioventricular discordant transposition of the great arteries may need surgery for the associated defects; thus VSD closure, relief of subpulmonary obstruction and tricuspid valve repair or replacement may be indicated for individual patients. Postoperative echocardiograms would therefore need to be individualized depending on the specific operation performed.



Long-axis view in congenitally corrected transposition of the great arteries. Note posterior pulmonary artery (PA). Subpulmonary stenosis with accessory atrioventricular tissue from both right and left atrioventricular valves. Ao, aorta.



Parasternal short-axis view of anterior levo-positioned aorta with posterior pulmonary artery bifurcation. Ao, aorta; PA, pulmonary artery.



Apical four-chamber view in congenitally corrected transposition of the great arteries. Dilated LA from tricuspid regurgitation and Ebstein's-like displacement of septal leaflet. The arrow indicates the level of tricuspid annulus. LA, left atrium; MV, mitral valve; TV, tricuspid valve.



Apical four-chamber view in congenitally corrected transposition of the great arteries demonstrating herniation of septal leaflet of tricuspid valve (TV) into ventricular septal defect. Arrow indicates septal leaflet of TV.



Chapter 19

Double-Outlet Right Ventricle

Double-Outlet Right Ventricle

This conotruncal malformation represents one type of ventriculoarterial alignment.

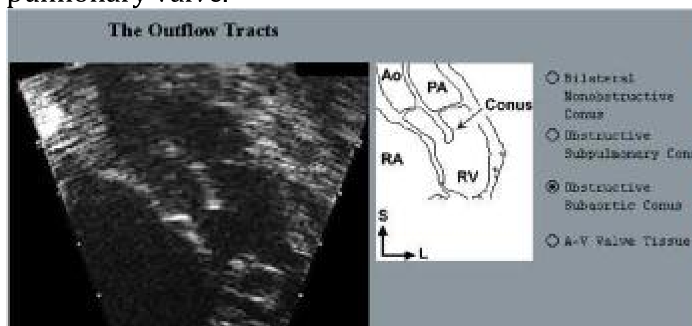
- Both great arteries arise primarily from the right ventricle.
- Variations in great arterial relationships, conal anatomy and outflow tract obstructions, the atrioventricular valves and associated septal defects lead to a wide spectrum of pathology and presentations.
- DORV maybe part of *visceroatrial heterotaxy syndrome*.(*randomization of cardiac, pulmonary, and gastrointestinal situs where coordinated signaling is absent. Such patients can have defects in almost all aspects of cardiogenesis. Often either the right or left side predominates with patients either having bilateral right-sidedness (asplenia syndrome) or bilateral left-sidedness (polysplenia syndrome*).

Associated Defects:

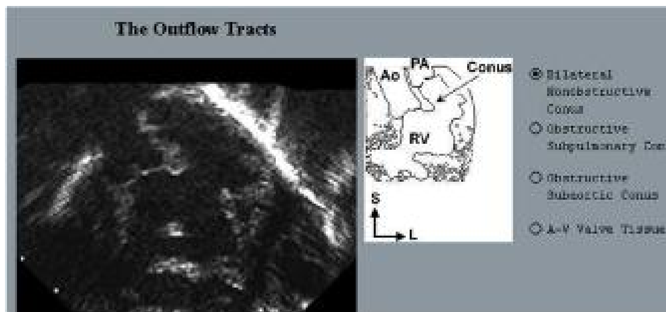
Great Arteries
VSD
Outflow Tracts

Great Arterial Relationships

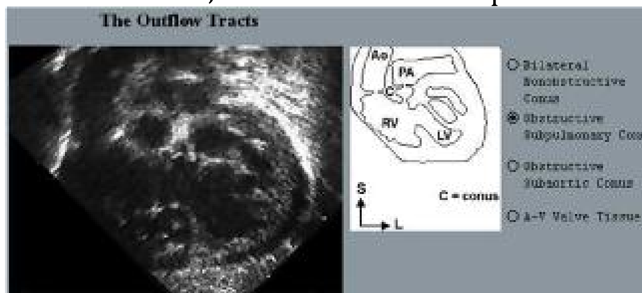
- Ø The spatial relationship of the aortic valve relative to the pulmonary valve is an important variable. Five types of relationships are possible:
- *D-malposition of the aortic valve*: Persistence of subaortic conus and absence of (or rudimentary) subpulmonary conus, with the aortic valve anterior and to the right of the pulmonary valve.



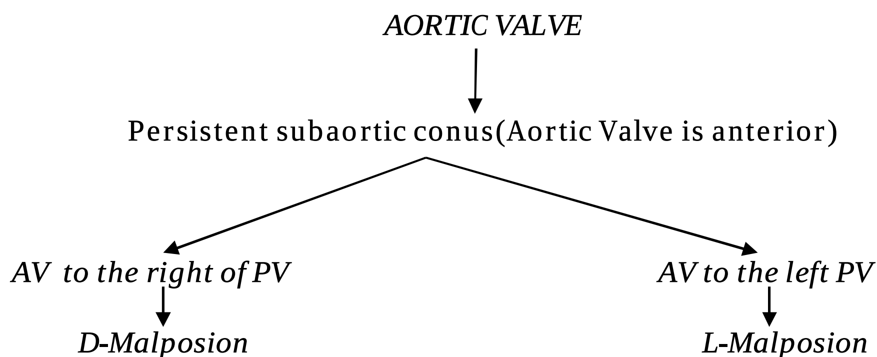
- *Side by side great arteries*: Persistence of subaortic and subpulmonary (bilateral) conus. The aortic valve is usually located to the right of the pulmonary valve in the same antero-posterior plane.



- *Solitus arrangement*: Persistence of subpuhnonary conus and absent (or rudimentary) subaortic conus, with the aortic valve posterior and to the right of the pulmonary valve.



- *Inverse arrangement*: Persistence of subpulmonary conus and absent (or rudimentary) subaortic conus, with the aortic valve posterior and to the left of the pulmonary valve. Usually seen in visceroatrial situs inversus.
- *L-malposition of the aortic valve*: Persistence of subaortic conus, usually absent or rudimentary subpuhnonary conus, with the aortic valve anterior and to the left of the pulmonary valve.



ECHO TIP:

- Best shown from subcostal long axis and parasternal sweeps.
- Conal anatomy and the spatial relationships of the aortic and pulmonary valves must be defined.
- There are no universally accepted definitions of a double outlet right ventricle. One school of thought, uses the '50%' rule, which defines the origin of a great, artery from a ventricle if > 50 % of the great artery arises over that ventricle, thus defining double outlet RV if the pulmonary artery arises from the right ventricle and > 50% of the aorta overrides the ventricular septum .
- A separate school defines the condition if blood exiting the left ventricle has to pass through the right ventricle (via a VSD) to enter either great artery.

Postop

The options for surgical repair of double outlet right ventricle depend on the type and severity of associated defects, especially outflow tract obstruction andVSD.

- A patient with a subaortic VSD and no outflow tract obstruction may need VSD closure to the aorta.
- With a subpulmonary VSD, the patient may need VSD closure to the pulmonary artery with an *arterial switch* operation.
- If there is pulmonary outflow obstruction, the *Rastelli repair* may be performed. This operation consists of closing the VSD to the aorta and placing a conduit (usually a pulmonary homograft) from the right ventricle to the pulmonary arterial confluence.
- Post operative echocardiograms should assess for conduit stenosis or regurgitation, branch pulmonary artery obstruction(esp. proximal) and include an estimate of RV pressure.
- Some patients with DORV may notbe candidates for biventricular repair, *e.g.*, with a noncommiited VSD which cannot be closed to either great artery. In such cases, staged repair for completion of the Fontan operation maybe needed.
- If there is associated A-V valve regurgitation or ventricular dysfunction, patients maybe precluded from completion of the Fontan operation, and may need cardiac transplantation.

Ventricular Septal Defects

- One critical issue in patients with DORV is the feasibility of biventricular repair, wherein VSD closure is achieved with a patch which directs left ventricular outflow into the aorta.
- Therefore, VSD location, size and relationship to the great arteries are critical issues. VSD location is variable; *definition of defect location must include a description of the relationship of the VSD to the great arteries*.
- *VSD locations* maybe *classified as* subaortic, subpulmonary (Taussig - Bing defect), doubly committed (related to both semilunar valves) or noncommitted (distant from both semilunar valves, *e.g.*, muscular or A-V canal type.
- *VSD size* is important; patients with a small interventricular communication or with far rightward malposition of the aortic valve who undergo defect closure may require VSD enlargement to avoid subaortic stenosis.
- *The relationship* of the VSD to the aortic valve determines the type of surgical procedure, *e.g.* patch closure for a patient with a subaortic VSD *versus* patch closure with arterial switch for a patient with a subpulmonary VSD.
- Patients with DORV may exhibt straddling of the atrioventricular valves across the ventricular septum. Since these A-V valve attachments may make biventricular repair difficult or impossible, they must be sought specifically in each case.

ECHO TIP:

- Best shown from subcostal and apical views.
- In order to identify structures that may impede future VSD closure to the aorta, *the VSD must be profiled from views that simultaneously show the aortic valve*. Structures that cross the VSD in these planes may complicate defect closure, and must be defined.
- In order to ensure that there are no A-V valve attachments straddling the ventricular septum, sweeps from multiple planes of interrogation at the level of the VSD must be used.
- In order to identify potential sources of postoperative outflow obstruction, preoperative echocardiograms must include a mental picture of the orientation of the VSD patch.
- Patients with a *subpulmonary VSD* (Taussig - Bing type of DORV) *may have associated left heart obstructive lesions including mitral stenosis*, *straddling mitral valve*, *subaortic stenosis and coarctation of the aorta*. These should be sought in each case.

Postop

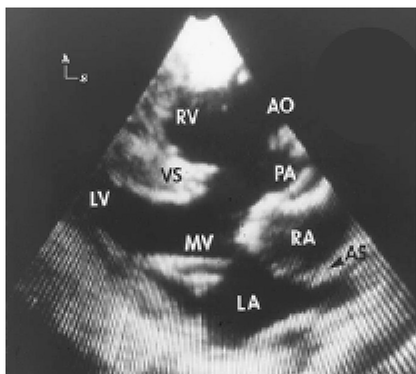
See above

The Outflow Tracts

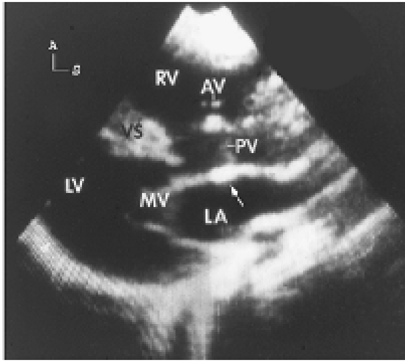
- Up to *50% of patients with DORV have pulmonary outflow tract obstruction*, usually due to deviation of the conal septum causing subpulmonary stenosis.
- Outflow tract obstruction can also be due to A-V valve tissue attachments, usually beneath the posterior great artery. Obstructions can occur at multiple levels.
- Thus, a patient with a subaortic defect and subpulmonary outflow obstruction may have pulmonary valvar, subvalvar and/or branch pulmonary artery stenosis.
- Similarly, apatient with a subpulmonary VSD (Taussig - Bing defect) may have mitral stenosis, parachute mitral valve, straddling mitral valve, subaortic stenosis, valvar aortic stenosis and/or coarctation of the aorta.

ECHO TIP:

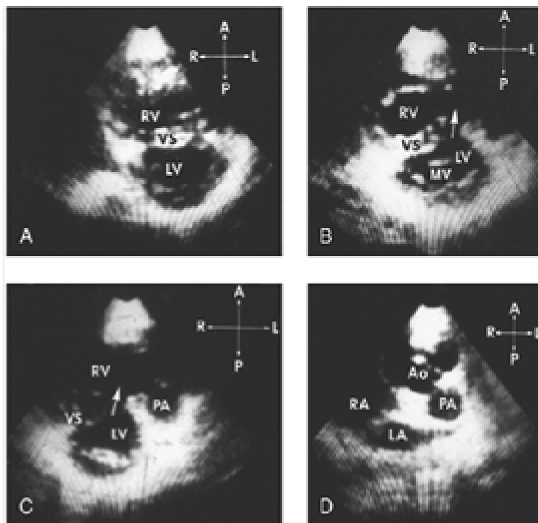
- Best shown from subcostal and parasternal views.
- Actual obstruction as well as the potential for future (postoperative) obstruction may be ruled out.
- In the presence of a VSD, anatomic definition of the substate for obstruction is more meaningful than the Doppler gradients.
- If one level of outflow tract obstruction is found, additional levels of obstruction may be sought.



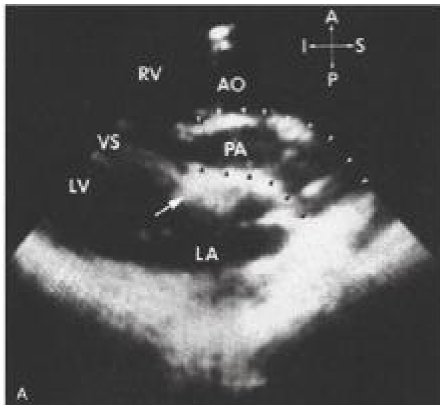
Parasternal long-axis scan in a patient with double-outlet right ventricle, with the aorta (Ao) anterior and to the left of the pulmonary artery (PA). Both great arteries are entirely committed to the right ventricular cavity and are observed in parallel orientation originating from the right ventricle (RV). In this standard long-axis scan, all four cardiac chambers and both great arteries are observed simultaneously. In this example, findings of left-juxtaposed atrial appendages are also evident. The right atrial (RA) appendage courses posterior to the great arteries to lie next to the left atrial appendage. The pulmonary valve appears slightly thickened, and in real time, the valve was dome shaped during systole, consistent with pulmonary stenosis. The mitral valve (MV) is markedly separated from the semilunar valves. AS, atrial septum; LA, left atrium; LV, left ventricle; TV, tricuspid valve; VS, ventricular septum



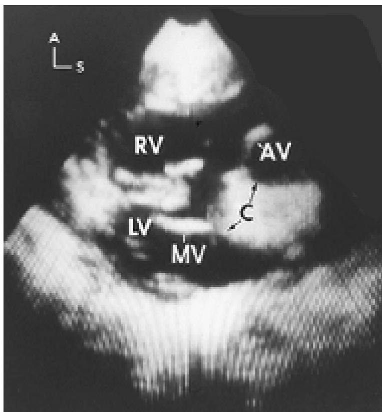
Parasternal long-axis scan in a patient with double-outlet right ventricle with the aorta anterior and right of the pulmonary artery with a subpulmonary VSD. There is a relatively small amount of conus tissue (arrow) separating the pulmonary valve (PV) from the mitral valve (MV). Both great arteries are entirely committed to the right ventricle (RV). AV, aortic valve; LA, left atrium; LV, left ventricle.



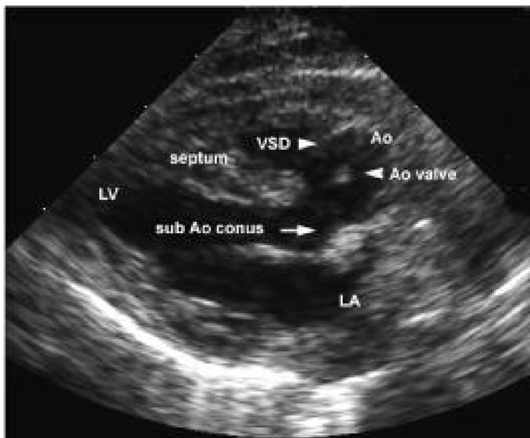
Short-axis scan of the heart from apex to base in a patient with double-outlet right ventricle. As the scan progresses up through the atrioventricular valves, a ventricular septal defect (VSD, arrow) is observed. At the cardiac base, it is apparent that the VSD is the only outlet from the left ventricular cavity and that both great arteries originate entirely from the anterior right ventricular cavity. The pulmonary artery (PA) is to the left and is separated from the VSD by a bridge of muscular tissue. Thus, the VSD is noted to be subaortic at the cardiac base. The aorta (Ao) is anterior and to the right of the PA, and a large conus is observed posterior to both great arteries. The pulmonary valve appears bicuspid and small in this patient with associated pulmonary stenosis. LA, left atrium; LV, left ventricle; MV, mitral valve; RA, right atrium; RV, right ventricle; VS, ventricular septum.



Parasternal long-axis scan, demonstrating posterior angulation of the pulmonary artery (PA) to the lungs, which allows its identification as the PA. Arrowheads indicate the posterior course of the PA. Both great arteries are entirely committed to the right ventricular cavity (RV). Subpulmonary conus is demonstrated by the large arrow.



Parasternal long-axis scan demonstrating double-outlet right ventricle. Large conus (C) separates the mitral valve (MV) from the aortic valve (AV), which originates entirely from the right ventricular (RV) cavity and is superiorly displaced by conus tissue. The ventricular septal defect (VSD) is the only outlet from the left ventricular cavity (LV) and is subaortic in position. VS, ventricular septum.



Long axis view. Note the large VSD which over-rides the septum. This case has a stenotic RV outflow tract but shows the sub-aortic conus between aortic valve and base of anterior mitral leaflet.

Chapter 20

Single Ventricle

Single Ventricle

- This condition exists when either the right ventricle or the left ventricle is absent .
- The term 'single ventricle' has also been used to describe the condition that exists when both AV valves or a common A-V valve open into one ventricular chamber.
- However, several fallacies implicit in this latter definition have been pointed out by Van Praagh et al.
- The definition of single ventricle used in this chapter refers to complete absence of one of the ventricles.
- *Presence of a hypoplastic ventricle is not considered to be the same as absence of that ventricle (although the latter may result in a functional single ventricle).*

Single Right Ventricle
Single Left Ventricle
Double-Inlet Left Ventricle with Normally Related Great Arteries (A-I Single Ventricle)
Double-Inlet Left Ventricle with Normally Related Great Arteries (A-I Single Ventricle)
Double-Inlet Left Ventricle with Left-Sided Subaortic Hypoplastic Right Ventricle (A-III Single Ventricle)
Double-Inlet Ventricle of Mixed Morphology
Double-Inlet Ventricle with Indeterminate or Undifferentiated Morphology
Single-Inlet Ventricle
Common-Inlet Ventricle

Single Right Ventricle

- This condition exists when the left ventricle is completely absent Echocardiographically, this is characterized by a *single trabeculated ventricular chamber*.
- If the left ventricle is absent, identification of the septal surface of the right ventricle may be difficult or impossible.
- The A-V valve morphology is variable, and may consist of either a common A-V valve (common-inlet RV) or two A-V valves (double-inlet RV). Both great arteries originate from the single right ventricle; there may be outflow tract obstruction of varying degree, due to varying causes.

ECHO TIP:

- Ventricular morphology is best seen from apical and subcostal views.
- The direction of ventriculair looping should be surmised from the contour of the ventricular septal surface; this may be difficult or impossible in a patient with a single right ventricle.
- By definition, the single ventricle provides systemic output Ventricular function and A-V valve *regurgitation must be addressed specifically*.
- The presence, location, severity and morphologic basis of *outflow tract obstruction* must be defined. If one level of outflow tract obstruction is detected, additional levels of obstruction

must be sought. Thus, if there is subaortic obstruction, the search for multilevel obstruction should *include assessment for coarctation*.

Postop

- The surgical option for patients with single ventricle is staged repair aimed at completion of the Fontan operation. If a patient is not a candidate for the Fontan operation, cardiac transplantation may be performed.

Single Left Ventricle

- This condition exists when the right ventricle is completely absent.
- Single left ventricle may occur with either d-Ioop or, more commonly, L-loop ventricles.
- It is characterized echocardiographically by a single ventricular chamber with a smooth septal surface. The septal surface of the single left ventricle separates it from the infundibulum, which is also termed the *'(rudimentary) outlet chamber'*. There is frequently a defect in the septum that separates the left ventricle from the infundibulum; this defect is termed the *bulboventricular foramen*.
- The A-V valve morphology is variable, and may consist of either a common A-V valve (common-inlet LV) or two A-V valves (double-inlet LV).
- The origin of the great arteries is variable. *Thus, either great artery or both great arteries may originate from either the left ventricle or the infundibulum*. If a great artery originates from the infundibular chamber, then the bulboventricular foramen is an important site of potential outflow tract obstruction; if there is restriction to outflow at this level, this is termed a restrictive bulboventricular foramen.

ECHO TIP:

- Ventricular morphology is best assessed from apical and subcostal views.
- The direction of ventricular looping should be surmised from the position of the ventricular septal surface. *Thus, if the septal surface is to the left of the left ventricle, the inference is that ventricular l-looping exists*.
- By definition, the single ventricle provides systemic output Ventricular function and *A-V valve regurgitation must be addressed specifically*.
- If one or both great arteries originate from the infundibular chamber, the diameter of the bulboventricular foramen must be measured in orthogonal planes. Available nomograms predict future restriction of the bulboventricular foramen if the area of the bulboventricular foramen is less than 2 cm^2/m^2 body surface area. Serial echocardio graphic evaluation is important in this situation because *progressive diminution* of the bulboventricular foramen can occur.
- The presence, location, severity and morphologic basis of outflow tract obstruction must be defined. If one level of outflow tract obstruction is detected, additional levels of obstruction must be sought. Thus, if there is subaortic obstruction, the search for multilevel obstruction should *include assessment for coarctation*.

Postop

The surgical option for patients with single ventricle is staged repair aimed at conpletionof theFontan operation.

Double-Inlet Left Ventricle with Normally Related Great Arteries (A-I Single Ventricle)

- Double-inlet left ventricle with a hypoplastic subpulmonary right ventricle and normally related great arteries is the classically described Holmes heart .
- This form of DILV remains relatively rare, observed in only 15% of the Van Praagh series .
- Embryologically, the VSD providing communication between the main left ventricular chamber and the hypoplastic right ventricle represents the primitive bulboventricular foramen.
- Frequently, the VSD creates significantly subpulmonary obstruction, resulting in a somewhat balanced circulation with some hypoxia and low pulmonary artery pressure.

Double-Inlet Left Ventricle with Right-Sided Hypoplastic Subaortic Right Ventricle (A-II Single Ventricle)

- Double-inlet left ventricle with right-sided hypoplastic subaortic right ventricle shares many anatomic features with a similar anomaly consisting of complete transposition of the great arteries with severe override and straddling of the right AV valve into the morphologic left ventricle and associated hypoplasia of the morphologic right ventricle .
- It was observed in 25% of the cases of single ventricle reviewed by Van Praagh et al.
- The embryologic development of various types of DILV representing the extreme forms of AV valve straddling easily supports the common features shared by these defects.
- In some cases, the AV valve morphology also may follow this schema, with the left-sided AV valve having morphologic features of a mitral valve and the right AV valve having features of a tricuspid valve.
- With ventricular and great artery discordance, the aorta also is typically right-anterior in location .
- Subaortic and pulmonary stenosis may occur.
- The conduction tissue enters the trabecular septum from an anterolateral node . The nonbranching bundle courses along the rightward rim of the VSD and subsequently distributes over the crest of the ventricular septum.

Double-Inlet Left Ventricle with Left-Sided Subaortic Hypoplastic Right Ventricle (A-III Single Ventricle)

- Double-inlet left ventricle with left-sided subaortic hypoplastic right ventricle *is the most common type of univentricular connection* . As the classically described type of single or primitive ventricle, it was encountered in 38% of the series review by Van Praagh et al..
- In this early series, most examples of this defect were thought to represent examples of ventricular l-looping with the morphologic left ventricle to the right and therefore with the morphologic mitral valve to the right of the morphologic tricuspid valve.
- Developmentally, one could speculate that this form of DILV shares anatomic features similar to AV and VA discordance (corrected transposition of the great arteries), but that severe overriding and straddling of the left-sided tricuspid valve ultimately resulted in predominant commitment of the left AV valve to the morphologic left ventricle and DILV .

- Thus, varying degrees of residual straddling of the left AV valve into the morphologic right ventricle would not be an unusual finding in this form of DILV.
- There is also significant variation in the size of the morphologic right ventricular cavity. It may be extremely hypoplastic with a tiny slitlike chamber or can be 75% or 80% of the size of a normal right ventricular chamber, particularly when the left AV valve straddles into the right ventricle.
- Various associated AV valve, semilunar valve, and outflow tract anomalies may occur in cases of DILV with a left-sided hypoplastic right ventricle.
- Subaortic obstruction is an important associated lesion that must be assessed preoperatively before considering a modified Fontan procedure.
- Significant ventricular hypertrophy substantially increases operative risk because of associated ventricular diastolic filling abnormalities and elevated left ventricular end-diastolic pressure.
- The conduction tissue abnormalities are similar to those described for AV and VA discordance . The AV node is anterolateral, and the nonbranching bundle courses into the ventricle anterior to the pulmonary outflow tract, subsequently coursing superiorly along the right (anterior) rim of the VSD.

Double-Inlet Ventricle of Mixed Morphology

- As a type of single ventricle without an outlet chamber, double-inlet ventricle of mixed morphology is a rare form of univentricular connection and occurred in only 5% of the series reported by Van Praagh et al. .
- Also called a common ventricle, it was designated by Van Praagh as the C type of single ventricle with absence of the ventricular septum or undivided ventricles with a rudimentary septum.
- A small apical ridge of ventricular septum often may separate the right and left ventricular zones of the heart .
- Relationships of the ventricular zones are usually consistent with normal ventricular locations or d-ventricular looping.
- Although some have dismissed this form as simply an extremely large VSD, functionally, there does not seem to be sufficient reason to exclude this type from being classified as a univentricular AV connection.
- AV valve straddling may occur, or there may be interdigitation of the AV valve tensor apparatus.
- Often, the great arteries are normally related; however, malposition with right- or left-anterior aorta may occur. Subvalvular and valvular pulmonary stenosis may be present.
- The conduction system has been described as normal, with a regular posterior position for the AV node .
- In some patients there may be dual AV nodes with a sling of conduction tissue connecting the two. The nonbranching bundle appears to descend into the remnant of ventricular septum that separates the right and left ventricular zones.

Double-Inlet Ventricle with Indeterminate or Undifferentiated Morphology

- Double-inlet ventricle with indeterminate or undifferentiated morphology is a form of single ventricle that often is considered to be a primitive form of univentricular AV connection *without a rudimentary chamber*. It shares many of the pathologic features of both double-inlet right ventricle and double-inlet of mixed morphology .

- This type of double-inlet ventricle often is diagnosed when no clear-cut differentiation or distinction of ventricular myocardium can be determined.
- Multiple coarse and irregular trabeculations may be present, providing chordae tendineae for both AV valves.
- Other portions of the ventricle may have smooth-walled components.
- AV valve abnormalities can include valvular stenosis or hypoplasia.
- Large, malformed AV valves often are regurgitant.
- The great arteries may be normally related. However, most commonly they are malposed with the aorta located anteriorly and to the right or left.
- Pulmonary stenosis with subvalvular and valvular stenosis or pulmonary atresia may be present.
- Location of the conduction system varies, with anterolateral and normally positioned posterior nodes described . The nonbranching bundle either penetrates directly into the right lateral wall of the ventricular chamber or descends through a large trabeculation toward the ventricular apex.

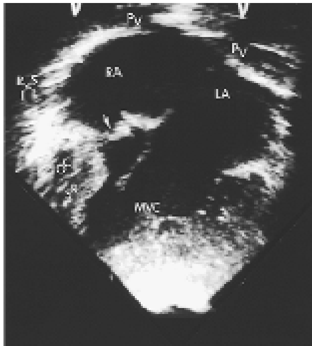
Single-Inlet Ventricle

- The common denominator of a single-inlet ventricle is a single AV connection to one ventricular chamber.
- It can include all forms of univentricular AV connection resulting from atresia or absence of either the right or left AV valve.
- The single-inlet connection may be to either a morphologic right ventricle with mitral valve atresia or a morphologic left ventricle with tricuspid valve atresia.
- AV concordance is common, but AV connection can be discordant.
- Similar to pathologic features observed with double-inlet ventricle, tricuspid valve atresia with single-inlet LV has a hypoplastic morphologic right ventricle that functions as an outlet chamber.
- With mitral valve atresia and single-inlet right ventricle, there is a hypoplastic rudimentary left ventricle, usually represented by a slitlike posterior chamber or trabecular mass of muscle.
- Ventricular and great artery concordance or discordance occurs with a range of great artery relationships from normal to transposition with a right or left anterior aorta.
- Subvalvular and valvular pulmonary stenosis can occur with concordant or discordant VA connections.
- Subaortic obstruction may occur predominantly with a restrictive VSD with discordant VA connections.

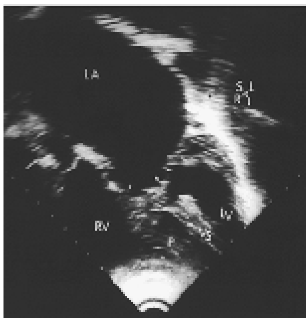
Common-Inlet Ventricle

- Common-inlet ventricle is a unique form of univentricular AV connection characterized by communication of both atria to a single-ventricular chamber by a common AV valve.
- It is unique by its *association with an AV septal defect* with absence of the primum portion, or, sometimes, the entire atrial septum.
- The usual form of this type of single ventricle is associated with situs ambiguous, particularly asplenia, and includes a common atrium, common AV valve, and common ventricle.

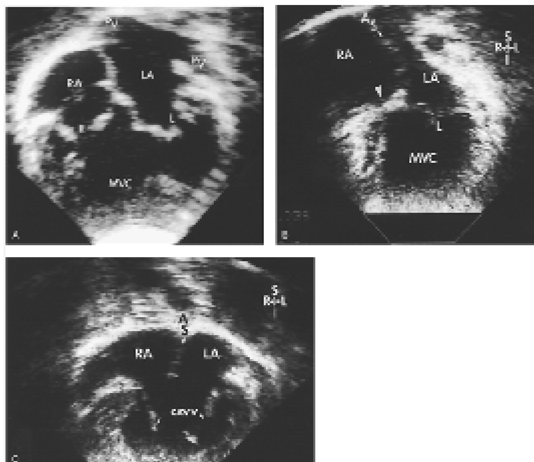
§ This form of complete AV septal defect often represents an extreme form of unbalanced AV septal defect, with the main ventricular chamber usually identified as a morphologic right ventricle with a severely hypoplastic morphologic left ventricle located posteriorly .



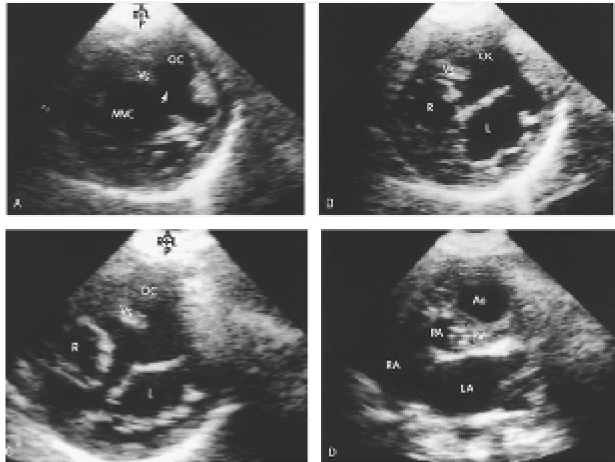
Double inlet with single atrioventricular (AV) valve. Echocardiogram demonstrating an apical four-chamber view. There is double-inlet left ventricle with atresia of the right AV valve. Note that the atretic right AV valve plate (arrow) is directed to the main ventricular chamber (MVC) of left ventricular morphology with malalignment of the atrial and ventricular septa. I, inferior; L, left; LA, left atrium; OC, outlet chamber of right ventricular morphology; PV, pulmonary vein; R, right; RA, right atrium; S, superior; SR, ventricular septal remnant.



Atrioventricular valve abnormalities. Echocardiogram demonstrating double-inlet right ventricle (RV) with severe stenosis of the left atrioventricular (AV) valve. There is a parachutelike deformity of the left AV valve with all the valve chordae committed to a single papillary muscle (P). The leaflets are also severely thickened (arrows). The left atrium (LA) is severely enlarged, consistent with severe left AV valve obstruction. A hypoplastic left ventricle (LV) is located posterior and to the left. I, inferior; R, right; S, superior; VS, ventricular septum.



Univentricular atrioventricular (AV) connections. Two-dimensional echocardiographic apical four-chamber views demonstrating (A) double-inlet, (B) single-inlet, and (C) common-inlet connections. In B, the arrowhead points out the AV junction. There is no malalignment of the atrial and ventricular septa. There is only the left AV valve committed to the main ventricular chamber (MVC). In C, there is a common inlet with a common AV valve (CAVV) draining both right and left atria. AS, atrial septum; L, left atrioventricular valve; LA, left atrium; PV, pulmonary vein; R, right atrioventricular valve; RA, right atrium.



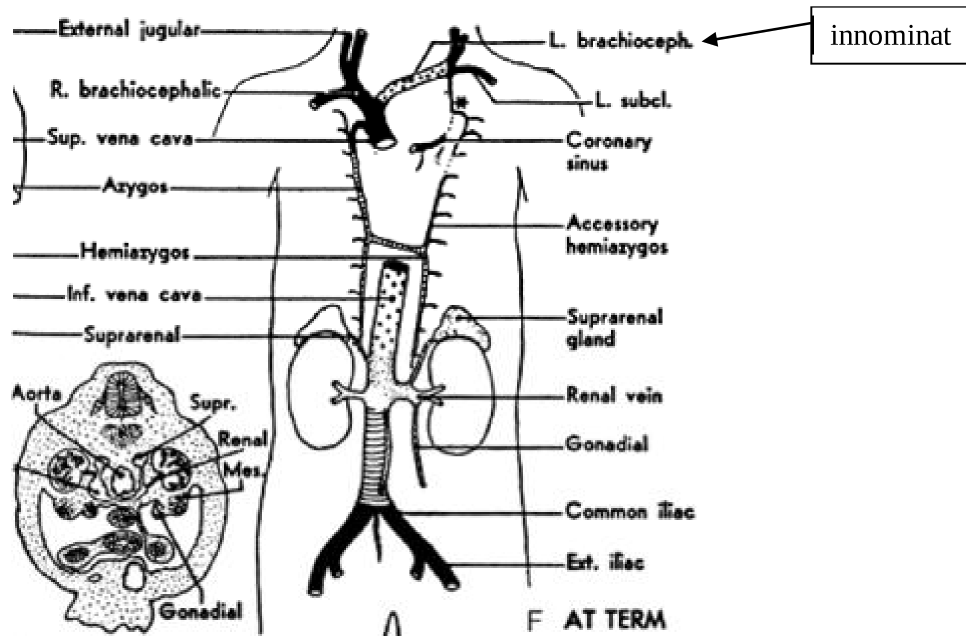
Progressive parasternal short-axis scans from apex (A) to base (D) in a patient with double-inlet left ventricle with left-anterior subaortic right ventricle. There is a large ventricular septal defect (white arrow) from the main ventricular chamber (MVC) of the left ventricular morphology into the hypoplastic outlet chamber (OC) of right ventricular morphology. The left (L) atrioventricular valve straddles into the right ventricular chamber through the ventricular septal defect. There is pulmonary valve (PV) stenosis with thickened leaflets (black arrows). Ao, aorta; LA, left atrium; PA, pulmonary artery; R, right atrioventricular valve; RA, right atrium; VS, ventricular septum.

Chapter 21

The Systemic Veins

The Systemic Veins

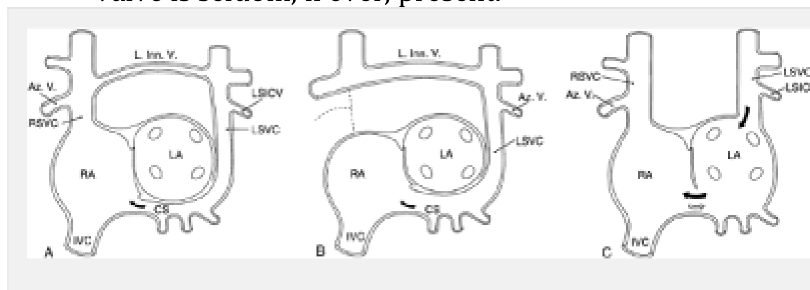
- The systemic veins are derived from multiple embryonic venous systems.
- The most primitive of these are the umbilical and viteltine veins, which develop into the inferior vena cava and hepatic veins.
- *Normal Development of the Right Superior Vena Cava and the Coronary Sinus* :
  - The right superior vena cava (SVC) extends from the confluence of the right and left innominate veins to the right atrium.
  - It is composed of the most proximal part of the right anterior cardinal vein and the right common cardinal vein.
  - The development of the left innominate vein at the 7th week of gestation usually is followed by the involution of the left SVC (LSVC), which becomes the ligament of Marshall .
  - As the transverse segment of the sinus venosus shifts rightward, it pulls the left horn of the sinus venosus along the posterior atrioventricular groove. The left horn of the sinus venosus and the adjacent part of the common cardinal vein receive the cardiac veins and form the coronary sinus.
  - The mode of formation of the coronary sinus is responsible for the following anatomic observations, which are helpful in making a diagnosis of certain congenital heart defects:
    - The orifice of a normally formed coronary sinus is always in the anatomically right atrium.
    - A persistent left SVC always continues with the coronary sinus since the left common cardinal vein is part of the coronary sinus and of the left SVC.
    - Functional connection (i.e., drainage) of a persistent LSVC or any other vein with the anatomically left atrium can occur only if the coronary sinus is partly or completely unroofed.
- Normal systemic venous development depends on a coordinated pattern of selective obliteration versus persistence of primitive venous channels. Any abnormality of the normal pattern may result in anomalies of systemic venous return. Identifying systemic venous anomalies may be of great importance, particularly if they result in a right-to-left shunt or if they occur in patients who are being staged for single ventricle (Fontan) repair.



Left Superior Vena Cava
Azygous Continuation of Inferior Vena Cava
Levoatriocardinal Vein

Left Superior Vena Cava

- The size of the LSVC varies. It may be smaller, equal, or larger than the size of the right superior vena cava (RSVC).
- A left innominate vein of variable size may be present in 60% of cases .
- The LSVC starts at the junction of the left jugular and left subclavian veins. It descends in front of the aortic arch and the left pulmonary vessels and, after receiving the left superior intercostal vein, it penetrates the pericardium. It then proceeds obliquely along the posterior wall of the left atrium and joins the coronary sinus in the posterior left atrioventricular groove .
- The flow of the LSVC blood into the coronary sinus results in its enlargement and posterior displacement of its orifice on the floor of the right atrium . The thebesian valve is seldom, if ever, present.



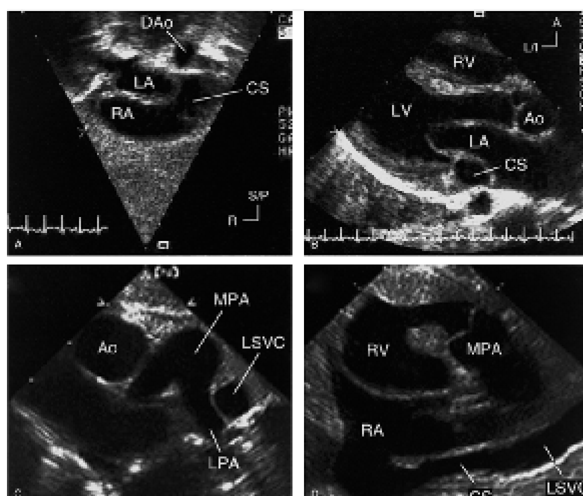
Persistent left superior vena cava (LSVC). A: LSVC drains via coronary sinus (CS) into right atrium (RA). The diameters of the LSVC and the left innominate vein (L. Inn. V.) vary inversely; the latter may be absent. B: Absence of the right superior vena cava. C: Bilateral superior venae cavae associated with unroofed coronary sinus and drainage of LSVC into left atrium (LA); the right atrial orifice of the

coronary sinus is enlarged and allows an interatrial communication. Az. V., azygos vein; IVC, inferior vena cava; LSICV, left superior intercostal vein; RSVC, right superior vena cava.

- This defect is due to *failure of normal resorption of the left anterior cardinal* vein. Most commonly, bilateral superior venae cavae are present. The left innominate (so-called bridging) vein may connect the two venae cavae; there is an inverse relationship between the size of the left innominate vein and the size of the left superior vena cava.
- The course of the left superior vena cava is *anterior to the aortic arch*, *left pulmonary veins and left pulmonary artery*. Approaching the heart, the LSVC courses rightwards and inferiorly, approaching the left posterior atrioventricular groove where it becomes the coronary sinus, which drains into the right atrium.
- Uncommonly, the coronary sinus maybe unroofed (i.e., lacking an anterior wall which normally separates it from the left atrium). When this occurs in a patient with drainage of a left superior vena cava to the coronary sinus, the LSVC will, in effect, drain into the left atrium.

ECHO TIP:

- *Imaging of a dilated coronary sinus is often the first clue to the diagnosis of a LSVC during the course of an echocardiographic examination*.
- The coronary sinus can be imaged from the subcostal, apical, and parasternal windows . It appears as a tubular structure in the posterior left atrioventricular groove with an opening into the posteroinferior aspect of the right atrium adjacent to the orifice of the IVC.
- The LSVC and its drainage into the coronary sinus can be imaged from the subcostal short-axis view in patients with adequate acoustic windows. In most patients, the LSVC can be imaged from the suprasternal notch or from the high left parasternal/subclavicular windows (Fig. below-D). From these windows, the presence and size of the left innominate vein can be imaged.
- In general, there is an inverse relationship between the caliber of the LSVC and the left innominate vein. Pulsed and color Doppler flow mapping are useful in demonstrating typical systemic venous flow patterns in the LSVC.
- Flow mapping by Doppler is also important in differentiating between a LSVC and other veins that may connect with the left innominate vein. These include partial or total anomalous pulmonary venous connection, a left superior intercostal vein, and a levoatrialcardinal vein. In contrast to a LSVC to an intact coronary sinus, however, the direction of blood flow in these veins is expected to be into the left innominate vein.



Echocardiographic features of persistent left superior vena cava (LSVC). A: Dilated coronary sinus (CS) imaged from the subcostal long-axis view. B: The dilated coronary sinus is seen in the posterior left atrioventricular groove from the parasternal long-axis view. C: The LSVC is seen anterior to the left pulmonary artery (LPA) in the parasternal short-axis view. D: The drainage of the LSVC to the coronary sinus (CS) and to the right atrium (RA) is seen from the high left parasternal sagittal view. A, anterior; Ao, aorta; DAo, descending aorta; LA, left atrium; L/I, left inferior; LV, left ventricle; MPA, main pulmonary artery; R, right; RV, right ventricle; S/P, superoposterior.

- Suspected based on a *dilated coronary sinus* seen from parasternal long axis view. The diagnosis is made by visualizing the drainage of the left superior vena cava into the coronary sinus *from suprasternal and subcostal views*.
- High frequency transducers and meticulous use of the transmit focus to allow fine focusing of the transducer are important for accurate definition of systemic venous anatomy.
- The presence or absence of a left innominate ("bridging') vein must be established.
- The presence or absence of a coronary sinus septum (anterior wall of coronary sinus wich normally separates the coronary sinus from the left atrium) must be defined, although this is very difficult .In the absence of the coronary sinus septum, the coronary sinus is unroofed, and drains directly to the left atrium.
- Rarely, the right superior vena cava maybe absent; this must be sought specifically.

Azygous Continuation of Inferior Vena Cava

- This condition is due to failure of the right subcardial vein to fuse with the hepatic vein.
- This results in agenesis of the inferior vena caval segment that normally connects the renal veins to the hepatic veins (suprarenal segment). The normally formed inferior portion of the inferior vena cava drains into the azygous vein, which ascends posteriorly and empties into the superior vena cava.
- This condition is frequently associated with visceroatrial heterotaxy.
- It is important to identify this condition prior to any cardiac catheterization, particularly a balloon atrial septostomy (catheter course would be unusual, making certain maneuvers difficult or impossible).
- Identification of this anomaly may also change the plans, timing and results of surgical anastomoses such as a bidirectional Glenn shunt.

ECHO TIP:

- The inferior (abdominal) portion of the azygous vein is *best shown from subcostal long axis view with rightward angulation*. The superior portion of the azygous vein is shown from *high right parasternal sagittal view*.
- Should be suspected if the inferior vena cava is not seen entering the right atrium in its usual position.
- The azygous vein is usually seen in *cross-section*, posterior to and to the right of the abdominal aorta.

Levoatriocardinal Vein

- This is a rare anomaly, characterized by a venous structure that connects the left atrium to either the superior vena cava or one of the innominate veins.
- The anomaly is typically seen in patients who have left atrial hypertension due to restriction to left atrial egress, such as mitral stenosis or atresia with an intact atrial septum (classically seen with hypoplastic left heart syndrome or its variants).

- The levoatriocardinal vein represents persistence of primitive systemicopulmonary venous connections that function as an avenue for left atrial decompression in these situations.
- This term is reserved for situations where the pulmonary veins return normally i.e., to the left atrium.

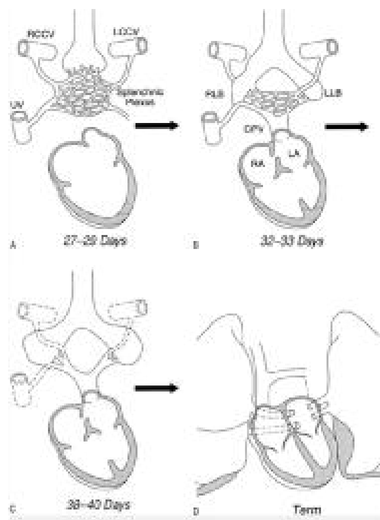
ECHO TIP:

- Ø Best shown from suprasternal and high left parasternal ('ductal') views.
- The drainage of each pulmonary vein must be established, since the diagnosis of a fevoatriocardinal vein depends on demonstrating normal pulmonary venous drainage to the left atrium.
- The cause of restriction to left atrial egress must be identified. Significant pulmonary venous hypertension may exist, and must be quantified.
- The course of the levoatriocardinal vein must be defined in its entirety, with particular attention paid to identifying the points of origin and drainage of this vein.

Chapter 22

The Pulmonary Veins

The Pulmonary Veins

- The pulmonary veins develop within the embryonic lung and initially connect to the splanchnic plexus, thus returning, in effect, to the systemic venous circuit.
- Eventually, an outpouching from the developing left atrium evaginates posteriorly and fuses with the pulmonary portion of the splanclmic plexus. This results in the normal state of connection of the pulmonary veins to the left atrium.
- Anomalies of pulmonary venous return are due to abnormal development of the common pulmonary vein.
- The specific pattern of abnormal pulmonary venous connection or drainage depends on the developmental stage at which the common pulmonary vein becomes stenotic or atretic.
- A primitive common pulmonary vein arises as an outpouching from the dorsal wall of the left atrium. With time, the common pulmonary vein communicates with the portion of the splanchnic plexus that drains blood flow from the lungs. Pulmonary venous connections to the cardinal and umbilicovitelline veins normally involute, and the common pulmonary vein becomes incorporated into the dorsal wall of the left atrium, ultimately typically giving rise to four separate pulmonary veins . Pulmonary venous developmental anomalies happen if any of these processes fails to occur properly. If the common pulmonary vein fails to connect to the splanchnic plexus and a splanchnic plexus communication with a cardinal or umbilicovitelline vein persists, some type of TAPVC or PAPVC will occur. If the common pulmonary vein fails to properly incorporate into the dorsal left atrial wall, pulmonary vein stenosis/atresia or cor triatriatum will occur .
- 

Development of the pulmonary veins. A: At 27 to 29 days of gestation, the primordial lung buds are enmeshed by the vascular plexus of the foregut (the splanchnic plexus). At this stage, there is no direct connection to the heart. Instead, there are multiple connections to the umbilicovitelline and cardinal venous systems. A small evagination can be seen in the posterior wall of the left atrium to the left of the developing septum secundum. B: By the end of the first month of gestation, the common pulmonary vein

establishes a connection between the pulmonary venous plexus and the sinoatrial portion of the heart. At this time, the connections between the pulmonary venous plexus and the splanchnic venous plexus are still patent. C: Next, the connections between the pulmonary venous plexus and the splanchnic venous plexus involute. D: The common pulmonary vein (CPV) incorporates into the left atrium so that the individual pulmonary veins connect separately and directly to the left atrium. LA, left atrium; LCCV, left common cardinal vein; LLB, left lung bud; RA, right atrium; RCCV, right common cardinal vein; RLB, right lung bud; UV, umbilical vein.

Supracardiac TAPVR
Intracardiac TAPVR
Infradiaphragmatic TAPVR
Partial Anomalous Pulmonary Venous Return

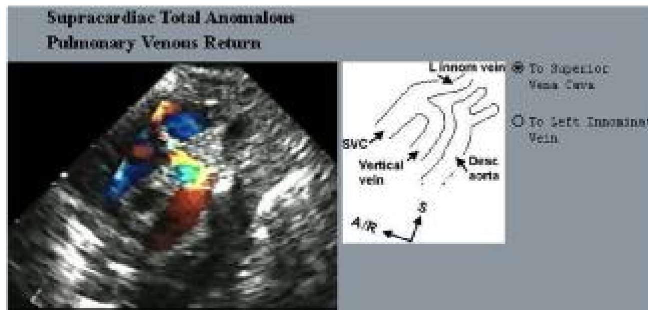
Supra cardiac Total Anomalous Pulmonary Venous Return

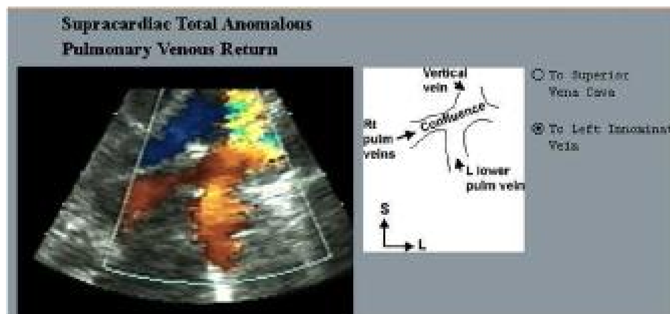
- This anomaly occurs due to *atresia of the common pulmonary vein* that occurs early in development, while the primitive venous channels that connect the splanchnic plexus to the systemic venous channels, ie., the right or the left cardinal veins, are still present and patent. Depending on whether the pulmonary veins connect to the right or the left sided cardinal system determines the pattern of drainage of the pulmonary veins.
- Supracardiac TAPVC is the result of failed development or atresia of the common pulmonary vein, which connects the embryonic lung buds with the left atrium.Collateral channels for pulmonary venous drainage develop via the fetal cardinal venous system. Usually, only 1 of these channels persists to become the vertical vein that connects the pulmonary venous confluence to the superior vena cava or the innominate vein.
- Typically, all the pulmonary veins meet in a confluence behind the left atrium, *and then connect to a supracardiac systemic vein via a vertical vein*. This vein may ascend rightwards behind the right pulmonary artery and the right mainstem bronchus, and drain into the superior vena cava or, rarely, into the azygous vein. Alternatively, the vertical vein may ascend leftwards, anterior to both the left pulmonary artery and the left mainstem bronchus (or between these two structures), draining into the left innominate vein.
- While obstruction is not a frequently reported finding in supracardiac TAPVR, multiple sites of obstruction are possible; thus, the entire vertical vein may be hypoplastic, or there maybe localized stenosis at the entrance of the vertical vein into the superior vena cava (or the left innominate vein). Even in the absence of obstruction, an adequate interatrial communication is essential for survival.
- Echocardiography should be able to define this diagnosis comprehensively in most cases.

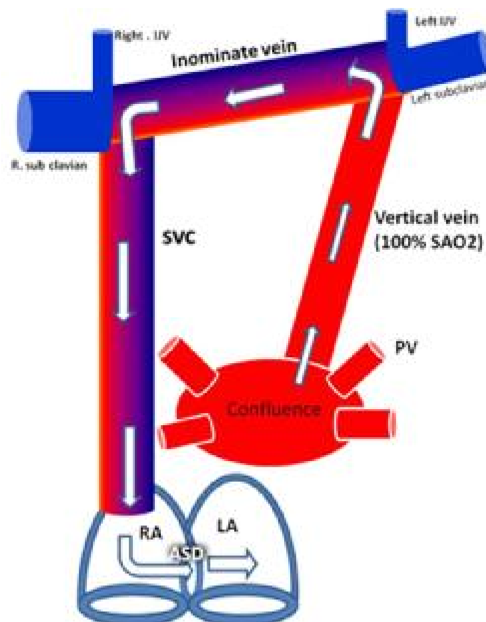
ECHO TIP:

- Best shown from suprasternal short axis and high right parasternal views.
- Each pulmonary vein must be identified and tracked using color flow Doppler (with a low Nyquist limit) to assess whether the vein itself is stenotic - and to confirm its drainage into the confluence.
- The diameter of the vertical vein must be defined along its entire length using 2-D imaging, and interrogated with pulsed wave Doppler to assess for any change in respiratory phasicity that may suggest obstruction.

- to ascertain that all four major pulmonary veins join the pulmonary venous confluence and that no additional pulmonary veins drain separately.
- image and determine the size of the pulmonary venous confluence and its relation to the left atrium.
- Particular attention should be paid to the entrance of the vertical vein into the superior vena cava or innominate vein as sites of potential obstruction.
- The location, size and direction of flow across any atrial septal defect should be assessed. *Right-to-left flow across the atrial septum should suggest the possibility of anomalous pulmonary venous return*.
- Right, ventricular systolic pressure and the degree of light ventricular pressure and volume overload should he assessed.

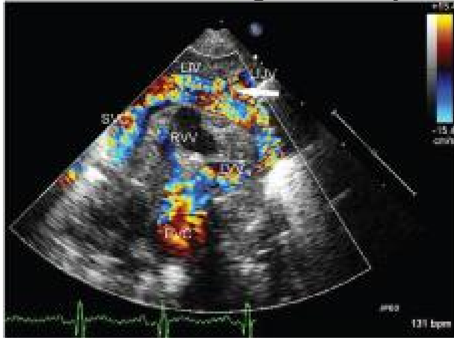




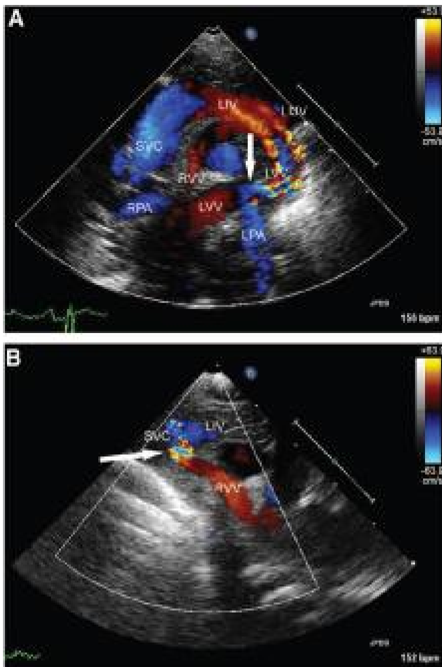


Postop:

- The repair of TAPVR consists of creation of an opening in the anterior wall of the pulmonary venous confluence and a complementary opening in the corresponding posterior wall of the left atrium; these two openings are anastomosed together, thus allowing free unobstructed normal return of pulmonary venous flow into the left atrium. The vertical vein is usually ligated at repair, and any interatrial communication is closed.
- Postoperative echocardiograms should assess for residual or recurrent pulmonary venous obstruction at the anastomotic site: individual pulmonary veins should also be assessed for obstruction. Right ventricular pressure should be estimated to assess for pulmonary hypertension. Serial echocardiograms are needed to assess for obstruction since this can be insidious, increasing in severity with linear growth.



Still frame in a high parasternal short axis. The left- and right-sided vertical veins are seen arising from the common confluence. The leftsided vertical vein is shown draining into the left innominate vein near the left internal jugular vein (arrow). The drainage site of the right-sided vertical vein into the left innominate vein is not shown. LIJV indicates left internal jugular vein; LIV, left innominate vein; LVV, left vertical vein; PVC, pulmonary venous confluence; RVV, right vertical vein; and SVC, superior vena cava.



A, High parasternal short-axis view of the left vertical vein, which courses from medio-postero-inferior to laterosupero- anterior. Doppler flow toward the transducer (from inferior to superior) is coded in red. Turbulent flow indicates compression of the vertical vein where it crosses over the left pulmonary artery (arrow). B, High parasternal long-axis vew of the right vertical vein, which is stenotic at the drainage site into the left innominate vein (arrow). LVV indicates left vertical vein; LIV, left innominate

vein; LPA, left pulmonary artery; RPA, right pulmonary artery; RVV, right vertical vein; and SVC, superior vena cava.

Intracardiac Total Anomalous Pulmonary Venous Return

- This anomaly occurs due to atresia of the common pulmonary vein that occurs early in development, while the primitive venous channels that connect the splanchnic plexus to the cardinal veins are still present and patent.
- Intracardiac TAPVR may *drain to the right atrium directly*, *or to the coronary sinus*; the latter pattern is due to connection of the pulmonary veins to the left cardinal system.
- Typically, all the pulmonary veins meet in a confluence behind the left atrium, and then connect to either the right atrium or the coronary sinus.
- Obstructed intracardiac TAPVR is rare; even in the absence of obstruction, an adequate interatrial communication is essential for survival.
- Echocardiography should be able to define this diagnosis comprehensively inmost cases.

ECHO TIP:

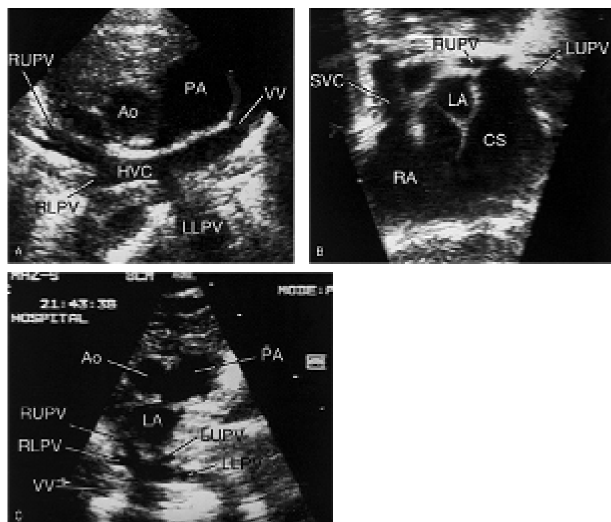
See above

Infradiaphragmatic TAPVR

- This anomaly occurs due to atresia of the common pulmonary vein that occurs early in development, while the primitive venous channels that connect the splanchnic plexus to the umbilicovitelline veins are still present and patent.
- *Typically*, *all the pulmonary veins meet in a confluence behind the left atrium*, which connects via a descending vertical vein which passes through the diaphragm to *connect to the portal vein*, *the ductus venosus*, *the inferior vena cava or a hepatic vein*.
- Obstructed Infradiaphragmatic TAPVR is the rule. The sites of obstruction within the vertical vein include the point of passage of the vein through the diaphragm, and the entrance of the vertical vein into the portal vein or other systemic vein. The length of the vertical vein and the passage of pulmonary venous return through the hepatic bed also impose resistance to pulmonary venous flow, which normally has alow velocity.
- Even with an adequate sized atrial septal defct, these patients are frequently critically ill, and a complete diagnosis must be established expeditiously; echocardiography should be able to define this diagnosis in most cases.

ECHO TIP:

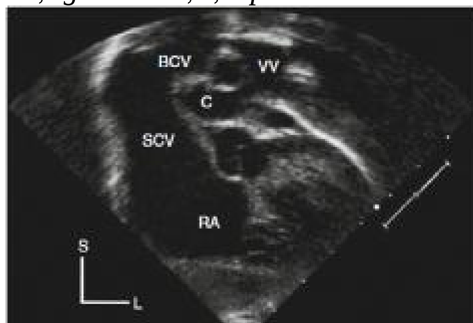
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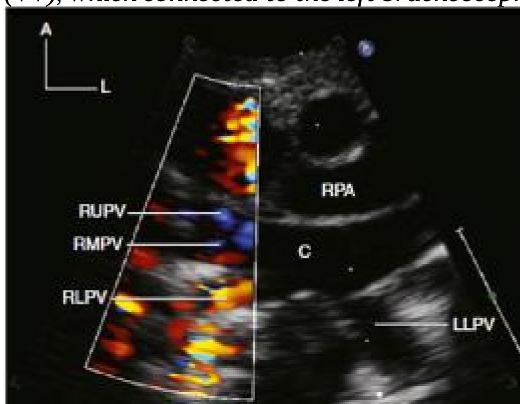
Echocardiographic delineation of totally anomalous pulmonary venous connection. A: High left parasternal transverse view of TAPVC to the left innominate vein. The right upper pulmonary vein (RUPV), right lower pulmonary vein (RLPV), and left lower pulmonary veins (LLPV) are seen joining a horizontal pulmonary venous confluence (HVC) behind the left atrium (the left upper pulmonary vein and the left atrium are not seen in this plane). The vertical vein (VV) ascends to the left of the main pulmonary artery (PA), which is dilated secondary to the volume load. Ao, aorta. B: Subcostal oblique view of totally anomalous pulmonary venous connection to the coronary sinus (CS), which is markedly dilated. Note the small size of the left atrium (LA). LUPV, left upper pulmonary vein; RA, right atrium; SVC, superior vena cava. C: High left parasternal transverse view of infradiaphragmatic TAPVC. From this view, the individual pulmonary veins can be seen as well as the proximal aspect of the descending vertical vein (VV).



In most cases of totally anomalous pulmonary venous connection, echocardiography reveals an echo-free non-pulsatile region beyond and clearly separate from the left atrium. In this image, taken from the subcostal long-axis view, the pulmonary venous confluence (C) can be seen posterior and superior to the left atrium (LA). Colour Doppler can be applied to this view, clearly distinguishing this vessel from a pulmonary artery. L, left; RA,right atrium; S, superior.



In this image, taken from the subcostal long-axis view angled posteriorly, note the severely dilated superior caval vein (SCV) emptyinginto the right atrium (RA), suggesting the presence of anomalous pulmonary venous connection. Further investigation revealed a confluence (C) behind the left atrium, leading to a vertical vein (VV), which connected to the left bracheocephalic vein (BCV). L, left; S, superior.



Suprasternal view of totally anomalous pulmonary venous connection. The appearance of the right pulmonary veins (RUPV, RMPV, and RLPV) entering into the confluence (C), along with the left lower pulmonary vein (LLPV) creates a cruciate appearance. This image, however, can mimic the normal appearance. Therefore, it is important to follow the course of the confluence (as is done in Fig. 24-20) A, anterior; L, left; RPA, right pulmonary artery.

Partial Anomalous Pulmonary Venous Return

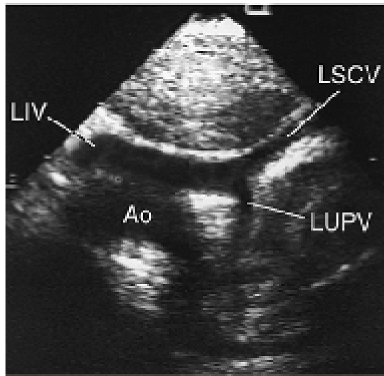
- This defect Is characterized by anomalous drainage of one or more (but not all) of the pulmonary veins to a systemic vein. An atrial septal defect is commonly associated. The left pulmonary veins most commonly drain to the left innominate vein. The right pulmonary veins may drain into the superior vena cava or the right atrium.
- Partial Anomalous Pulmonary Venous Connection with Intact Atrial Septum.
  When a single pulmonary vein is anomalously connected, the anomalously draining blood flow is about 20% to 25% of the total pulmonary blood flow . It is of such slight hemodynamic significance that the lesion is rarely recognized clinically.
- Partial anomalous pulmonary venous connection or drainage has been seen in association with other congenital cardiac defects and syndromes. Most notably, patients with visceral heterotaxy and polysplenia have a high incidence of PAPVD secondary to malposition of the septum primum, and patients with asplenia have a high incidence of TAPVC.
- A rare but clinically important association is that of anomalous pulmonary venous connections with tetralogy of Fallot.

ECHP TIP:

See above

Postop

The repair of partial anomalous pulmonary venous return consists of re-routing pulmonary venous return to the left atrium. The specifics of the repair depend upon the pattern of anomalous drainage in a given case. Thus, a sinus venosus atrial septal defect is closed with a baffle that redirects the anomalous right-sided pulmonary veins to the left atrium. Anomalous drainage of the left-sided pulmonary veins is treated, when possible, by direct anastomosis of the vertical vein or 'confluence' to the left atrium.



Echocardiogram from the high left parasternal window in the transverse view showing partial anomalous pulmonary venous connection of the left upper pulmonary vein (LUPV) to the left innominate vein (LIV). Ao, aorta; LSCV, left subclavian vein.

Chapter 23

Cardiac Tumors

Cardiac Tumors

- Primary cardiac tumours are most frequenty benign lesions. They may originate from endocardium (e.g., myxoma), myocardium (e.g., rhabdomyoma) or pericardium (e.g., mesothelioma).
- The commonest cardiac tumors encountered in children are rhabdomyomas, followed by fibromas and myxomas.
- Tumors maybe either single or multiple, and either circumscribed or diffusely infiltrating into adjacent myocardium. They may be pedunculated or sessile, protruding into one or more of the cardiac chambers, obstructing inflows or outflows, or interfering with A-V valve or semilunar valve function.
- Secondary (metastatic) cardiac tumors may involve myocardium and/or pericardium. These may spread to the heart from the primary focus in a viscus (e.g., liver or kidney) via the systemic veins. Hemorrhagic pericardial effusions may result from minor metastases into the pericardium.
- Echocardiography is the method of choice for diagnosis and complete delineation of tumors.

ECHO TIP:

- Since tumor location is variable, visualizing tumors requires a comprehensive study with crisp structural definition using high-frequency 2-D imaging from all echocardiographic views.
- If one tumor is seen, additional tumors must be sought.
- Tumor echogenicity maybe a clue to the type of tumor. For example, rhabdomyomas are *typically highly echodense*, *nodular and well-circumscribed masses*.
- Tumor size, shape and margins must be defined. This is important for purposes of serial study as well as for surgical planning, to determine the feasibility and extent of tumor resection.
- The degree of functional impairment attributable to the tumor must be assessed. Thus, the function of A-V valves and semilunar valves, atrial and ventricular filling and emptying should each be assessed.
- It may b e difficult or impossible to identify the edges of diffusely infiltrating tumors by echocardiography. Additional modes of imaging, including MRI studies, maybe needed.

Chapter 24

Vegetations & Thrombi

Vegetations & Thrombi

- Intracardiac thrombi (clots) typically occur inpatients with low-flow states due to depressed ventricular function, classically with dilated cardiomyopathy.
- Thrombi have a predilection for certain typical locations which exhibit decreased wall motion, *typically the atrial appendages and the ventricular apices*.
- Other situations that involve a high risk for developing intracardiac thrombi include prosthetic valves and indwelling lines.
- Thrombi that occur on cardiac valves are generally *called vegetations*. These may be non-infectious or may occur in the setting of infectious endocarditis.
- Patients with structural heart disease associated with turbulent, high-velocity flow are at particular risk for developing infectious endocarditis with intracardiac vegetations.
- The most common location of vegetations is *downstream along the pressure gradient*.
- Vegetations maybe single or multiple, and maybe either mobile or firmly attached to underlying endocardium or valve structure.
- Rarely, a patient with a structurally normal heart may develop endocarditis with vegetations. The typical scenario is an immunocompromised patient with an indwelling central venous catheter, who may have vegetations at the tip of the catheter.
- *Echocardiography cannot help in determining whether an echodense intracardiac mass is a thrombus or an infected vegetation; the clinical setting is more helpful in this regard*.

ECHO TIP:

- Since vegetations may occur anywhere in the heart, a comprehensive study is mandated with crisp structural definition using high-frequency 2-D imaging from all echocardiographic views.
- In patients with central venous lines, multiple planes of interrogation including high right parasternal sagittal views should be employed. Any occlusion or obstruction of systemic veins should also be sought.
- Typical sites of predilection, including sites of turbulent flow, A-V and semilunar valves, aortopulmonary shunts, venous catheter tips, the atrial appendages and ventricular apices should be examined specifically. If one intracardiac mass is seen, additional masses must be sought.
- Mass location, size and mobility must be determined by echocardiography.
- The degree of functional impairment attributable to the mass including function of A-V valves and semilunar valves, ventricular size and function must be assessed.
- Performing serial evaluations is important to monitor for change in size, location and mobility of these masses.
- It is particularly challenging to diagnose masses within the atrial appendages, due to the normal corrugations of the pectinate muscles. Transesophageal echocardiography may be needed for definitive diagnosis.

Chapter 25

The Pericardium and Pleura

The Pericardium and Pleura

The pericardium and the pleura are bi-layered membranes that invest the heart and lungs respectively. The importance of these membranes to the echocardiographer lies in the possibility of fluid collection between the two layers, manifesting as pericardial and pleural effusions respectively.

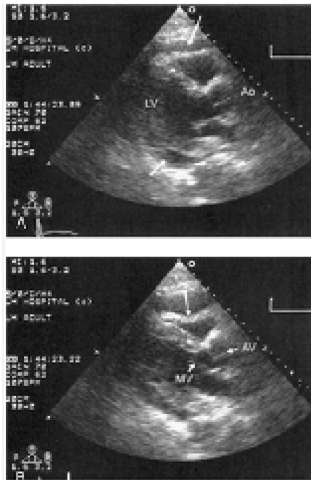
Pericardial effusion
Pleural effusion

Pericardial Effusion

- Pericardial effusions may develop secondary to viral pericarditis or in the postoperative period after cardiac surgery, due to either hemorrhage or postpericardiotomy syndrome. Rarely, bacterial infections may cause purulent pericardial fluid collections.
- An effusion is seen echocardiographically as an echofree space immediately outside and adjacent to the epcardial surface of the cardiac chambers. This may be shown by M-mode as well as by 2-D imaging.

ECHO TIP:

- Ø Best shown from a combination of parasternal and subcostal views.
- The size of the effusion should be measured at end-diastole, at several points around the circumference of the heart.
- If the effusion is large, diastolic collapse of the atria (and, in severe cases, the ventricles) is evident; in equivocal situations, this should be sought using M-mode echocardiography and the EKG tracing on the echo machine. This finding suggests actual or impending cardiac tamponade which is a medical emergency requiring drainage of pericardial fluid (pericardiocentesis).
- Echocardiography guidance improves the safety of pericardiocentesis. Imaging should be adequate to assess the distance of the fluid collection from the site of needle entry, location of the needle tip, its proximity to the cardiac chambers and the adequacy of the pericardiocentesis procedure (i.e., any residual fluid).



Two-dimensional echocardiographic images of pericardial tamponade. A: Parasternal long-axis view in systole. B: Diastolic collapse of the right ventricular free wall (anterior arrow). Ao, aorta; AV, aortic valve; LV, left ventricle; MV, mitral valve.

Pleural Effusion

- Pleural effusions may develop secondary to viral pericarditis or in the postoperative period after cardiac surgery, due to either hemorrhage or post-pericardiotomy syndrome. Rarely, bacterial infections may cause purulent fluid collections in the pleural space.
- An effusion is seen echocardiographically as an echofree space.

ECHO TIP:

- Best shown from subcostal views.
- Echocardiography can demonstrate medium-to-large sized pleural effusions.
- With large effusions, the collapsed lung may be seen compressed against the mediastinun, which may be pushed over to the opposite hemothorax.
- However, echocardiography cannot reliably quantify pleural effusions.

Differential diagnosis of pericardial and pleural effusions

Pericardial	Pleural
Ends anterior to the descending aorta	Ends posterior to the descending aorta
Minor overlapping of LA	May significantly overlap LA
Fluid between heart and diaphragm on subcostal view	No fluid between heart and diaphragm on subcostal view
Tamponade may be present	No signs of tamponade
May be >4 cm	Rarely >4 cm
If large, swinging of the heart	Heart fixed

Echocardiographic evidence of tamponade

- Dilated IVC (>2 cm) with inspiratory collapse of <50% .
- Fall in aortic or early diastolic mitral velocity during inspiration >25%.

- Prolonged and widespread diastolic RV collapse
- *Note that collapse of the RA and RV outflow tract are non-specific*

Signs.

Chapter 26

Cardiomyopathies

Cardiomyopathies

- The term defines disorders of myocardium that may be primary (unassociated with structural or systemic disease) or secondary (related to a systemic disease).
- Cardiomyopathies are classified based on the pattern of myocardial involvement; the three major types are dilated (congestive), hypertrophic and restrictive.
- The echocardiographic diagnosis of cardiomyopathy consists of definition of the type of myocardial involvement and ruling out a structural basis for myocardial abnormality (e.g., coarctation of the aorta as a potential cause of a 'dilated cardiomyopathy - like' picture).

Dilated
Restrictive
Hypertrophic
Non-compaction of Myocardium Cardiomyopathy

Dilated Cardiomyopathy

- This condition is characterized by left ventricular systolic *(with or without associated diastolic)* dysfunction and dilatation. Depressed myocardial contractility leads to low cardiac output.
- The etiology of dilated cardiomyopathy is unknown; the condition is probably one of the sequelae of viral myocarditis in most cases.
- Familial occurence of dilated cardiomyopathy and inborn errors of metabolism, including glycogen storage disease and mitochondrial abnormalities, are uncommon causes of this condition.
- Endocardial fibroelastosis also most often presents as a dilated cardiomyopathy.
- Dilated cardiomyopathy is characterized by a dilated, poorly contractile left ventricle, with akinetic or dyskinetic areas.
- A variable degree of mitral regurgitation may be present due to dilation of the mitral anulus.
- Spontaneous echo contrast maybe evident in severe cases, with echo dense masses (thrombi) in the atrial appendages and/or within ventricular apical trabeculations.

ECHO TIP:

- All echocardiographic views should be used to define wall motion, since regional wall motion abnormalities are common.
- Asymmetric left ventricular wall motion invalidates the geometric assumptions implicit in calculating fractional shortening; therefore, left ventricular ejection fraction should be calculated in such cases.
- Serial echocardiography evaluation is critical for determining clinical course.
- The presence and severity of mitral regurgitation should be assessed.
- The *peak velocity of tricuspid regurgitation* should be measured to enable calculation of right ventricular systolic pressure.
- The left atrial appendage and left ventricular apex should be examined for any echodense masses, which would suggest thrombi in this clinical setting.

- *Increased echodensity* of left ventricular endocardium may suggest *endocardial fibroelastosis*, and should be noted.
- Anomalous origin of the left coronary artery can lead to left ventricular dilatation and dysfunction that resemble dilated cardiomyopathy. *Therefore, in every case of dilated cardiomyopathy, the origins of the coronary arteries must be shown using high-frequency transducers*. This must be confirmed by demonstrating prograde color flow in both coronary arteries.
- Checklist for reporting LV dilatation
  1. LV dimensions, including wall thickness
  2. LV systolic and diastolic function
  3. RV size and function
  4. Pulmonary pressure
  5. Valve function
  6. Thrombus?

Postop

- Surgical options for patients with cardiomyopathy are limited. The definitive treatment is cardiac transplantation; for details on the post-transplant heart, please go to Chapter 30.
- Patients with hypertrophic cardiomyopathy and severe left ventricular outflow tract obstruction due to asymmetric septal hypertrophy may bepalliated with a septal myomectomy.
- Serial postoperative echocardiograms should assess for recurrent or residual left ventricular outflow obstruction and for new or increased mitral regurgitation.

Restrictive Cardiomyopathy

- This rare condition is characterized by decreased ventricular compliance in the face of normal ventricular size and function.
- The echocardiography (2-D) manifestations of restrictive cardiomyopathy are *massive atrial enlargement in the absence of significant A-V valve dysfunction*. This condition usually leads to *rapid and severe pulmonary hypertension*.

ECHO TIP:

- Ø Best shown from parasternal and apical views.
- *Massive biatrial dilatation* is the *hallmark* of this condition.
- Decreased ventricular compliance leads to rapid ventricular filling in early diastole, leading to the classic Doppler findings of attenuation of the late diastolic phase of ventricular filling. This is seen as an increased peak E/A ratios with shortened A- V valve deceleration times.
- Measurement of atrial size is important for serial comparisons.
- *Pulmonary hypertension* must be sought diligently, since it occurs with high frequency and severity in this disease.

Postop

See above

Hypertrophic Cardiomyopathy

- This term refers to left ventricular, and *often biventricular myocardial hypertrophy* in the absence of structural basis for hypertrophy.

- This may be primary (idiopathic) or, less commonly, secondary to inborn errors of metabolism such as glycogen storage disease or mitochondrial myopathy.
- Concentric or asymmetric (typically septal) hypertrophy may be seen; the severity of hypertrophy can change dramatically with time.
- The mitral valve is frequently abnormal, and systolic anterior motion (SAM) of the anterior mitral leaflet is a classic finding. This abnormal motion of the anterior mitral leaflet contacts the hypertrophic basal ventricular septum during ventricular systole, leading to left ventricular outflow tract obstruction.
- *Basal septal hypertrophy and SAM* together lead to endocardial thickening and a 'contact lesion'in the left ventricular outflow tract.
- Abnormal motion of the anterior mitral leaflet leads to imperfect systolic mitral leaflet coaptation resulting in mitral regurgitation.
- *Left ventricular diastolic dysfunction* is considered a part of this disease complex in adults; it is currently difficult to diagnose in children since there are no standard age-specific criteria for diagnosis of diastolic dysfunction.

ECHO TIP:

- Ø Best seen from parasternal long axis and apical views.
- The extent of septal hypertrophy (concentric or asymmetric) must be assessed.
- Measurements of end-diastolic septal diameter at several locations including the point of maximum diameter *are* important for purposes of serial echocardiographic evaluation.
- Systolic anterior motion of the anterior mitral leaflet should be shown using slow speed playback and freeze-frame.
- The presence, severity and morphologic basis of left ventricular outflow tract obstruction must be identified.
- The presence and severity of mitral regurgitation must be assessed.
- Left ventricular systolic function must be assessed, since left ventricular systolic dysfunction can occur in severe cases.
- Assessment of diastolic function may be difficult particularly in children with rapid heart rates. Peak E/A ratio reversal is an indicator of diastolic dysfunction.

Postop

See above

Non-compaction of Myocardium Cardiomyopathy

- Non-compaction of the ventricular myocardium ("spongy myocardium") is a rare congenital cardiomyopathy of children and adults resulting from arrested myocardial development during embryogenesis.
- Prior to formation of the epicardial coronary circulation at about 8 weeks of life, the myocardium is a meshwork of interwoven myocardial fibers that form trabeculae and deep trabecular recesses.
- That increased surface area permits perfusion of the myocardium by direct communication with the left ventricular cavity.
- Normally,as the myocardium undergoes gradual compaction, the epicardial coronary vessels form.
- In this disorder, echocardiography demonstrates a thin epicardium with extremely hypertrophied endocardium and prominent trabeculations with deep recesses. These features tend to be apically localized since compaction would normally proceed from base to apex, and from epicardium to endocardium.

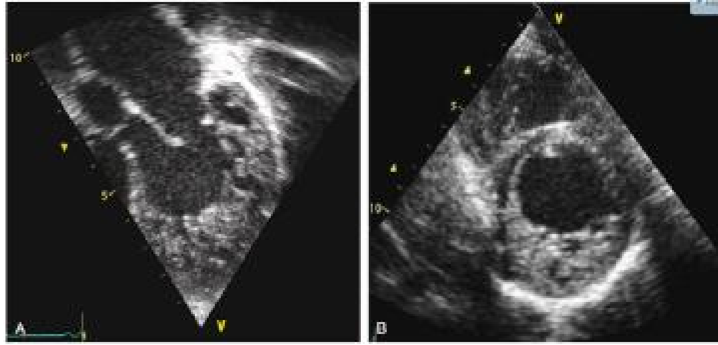
- Clinical presentation consists of congestive heart failure with depressed left ventricular systolic function, ventricular arrhythmias, arterial thromboemboli from thrombus formation within the inter - trabecular recesses, as well as restrictive physiology from endocardial fibrosis.

ECHO TIP:

- Diagnosis can be made using cross sectional echocardiography .
- Several diagnostic criterions have been proposed.
  - One suggests a ratio of 0.5 or less for the distance from the epicardial surface to the trough of a trabecular recess compared to the distance from the epicardial surface to the peak of the trabeculation measured in end- diastole.
  - Another requires the presence of numerous, excessively prominent trabeculations and deep intertrabecular recesses supplied by blood on colour Doppler, and a bilayered arrangement with a thin compacted layer and a thick non-compacted layer, the ratio of noncompacted to compacted endocardial layers being greater than 2 when measured in end-systole, in the absence of coexisting structural cardiac abnormalities.
  - A third requires the presence of more than three trabeculations protruding from the left ventricular wall distal to the papillary muscles visible in one imaging plane and associated with intertrabecular spaces visualised using colour Doppler.
- Each proposed criterion has limitations, and the three are probably complementary in making the diagnosis.
- Although awareness of the condition is increasing, some patients may be misdiagnosed as having distal or apical hypertrophic cardiomyopathy,and an overlap with dilated cardiomyopathy is also recognised.
- Alternative imaging modalities, particularly cardiac magnetic resonance imaging, may prove useful for distinguishing between left ventricular non-compaction and hypertrophic cardiomyopathy.
- In some children, an undulating phenotype characterised by alternating features of dilated cardiomyopathy and hypertrophic cardiomyopathy has been reported.



Transthoracic short axis view showing prominent trabeculations and deep intertrabecular recesses localized to the anterior and antero-lateral segments.



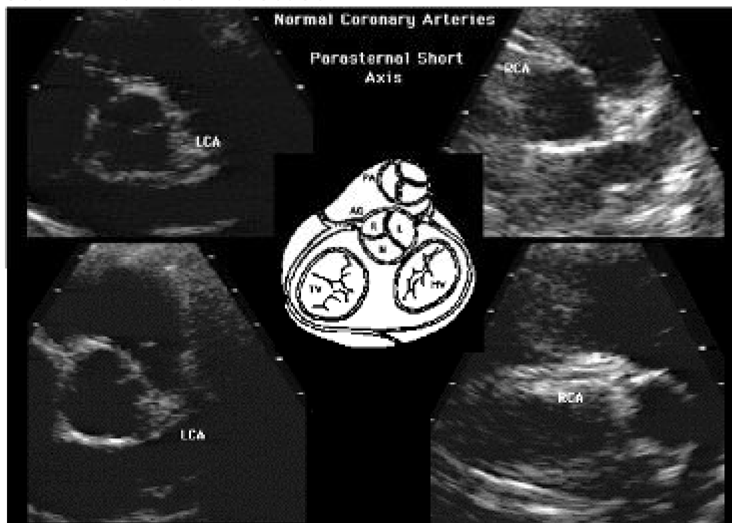
Echocardiographic features of left ventricular non-compaction. A is an apical five-chamber frame from a child with left ventricular non-compaction.
It shows left ventricular trabeculations and deep recesses extending into the left ventricular mid-cavity. B is a parasternal short-axis view from another child with left ventricular non-compaction, showing a hypertrabeculated area in the distal portion of the left ventricle.

Chapter 27

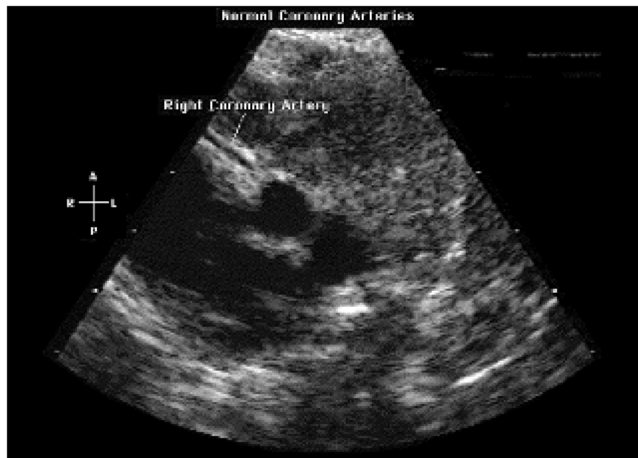
The Coronary Arteries

The Coronary Arteries

- The coronary arteries normally develop as evaginations from the aortic root extending into the myocardium to establish connection with intramyocardial sinusoids.
- In the normal two coronary artery system, the left main coronary artery and the right coronary artery arise form the respective sinuses of Valsalva.
- The left main coronary artery bifurcates into the left anterior descending and the circumflex coronary arteries.
- The left anterior descending coronary artery runs anteriorly and inferiorly in the anterior interventricular sulcus, while the circumflex coronary artery runs posteriorly in the left atrioventricular groove.
- In the more common right dominant coronary circulation, the left coronary artery supplies the anterior half of the ventricular septum and most of the left ventricle. The right coronary artery supplies the right ventricle, posterior half of the ventricular septum (via the posterior descending coronary artery) and the inferior walls of the left ventricle. In a left dominant circulation, the entire left ventricle and ventricular septum are supplied by the left coronary artery.
- Congenital anomalies of the coronary arteries may relate to abnormal origin or course, or fistulous communications with the right heart.
- Acquired coronary artery disorders that are seen in children include aneurysms, typically seen in Kawasaki disease.



Normal coronary arteries.



Right Coronary Artery

Anomalous Left Coronary Artery (ALCA)
Coronary Artery Aneurysms

Anomalous Origin of Left Coronary Artery from Main Pulmonary Artery

- This anomaly is due to either abnormal septation of the conotruncus into aorta and pulmonary artery, or due to persistence of the pulmonary buds with involution of the aortic buds that eventually form the coronary arteries.
- The clinical manifestations of congestive heart failure are caused by left ventricular ischemia and infarction due to perfusion of the left coronary artery by deoxygenated blood at low perfusion pressure.
- There is a coronary steal phenomenon due to collateral flow via the right coronary artery into the pulmonary artery.

ECHO TIP:

- Ø Best shown from angulated parasternal short axis views.
- The diagnosis requires tie demonstration of the origin of the left main coronary artery from the main pulmonary artery, with retrograde filling of the left main coronary artery.
- *Dilation of the right coronary artery is common*, and intramyocardial coronary arterial collaterals maybe seen.
- Dilation, dysfunction, echodensity and dyskinesis of the left ventricular walls, as well as regional wall motion abnormalities and mitral regurgitation due to papillary muscle dysfunction may be seen.
- The length of the left main coronary artery prior to its bifurcation, and the location of its insertion into the main pulmonary artery should be established.

POST OP:

- Surgery consists of reimplantation of the origin of the left main coronary artery into the aortic root.
- In rare situations where the coronary artery anatomy does not allow this partial switch, an aortopulmonary window may be created followed by fasluoning of a tunnel from the left main coronary artery to the aorta.

- Postoperative echocardiograms should assess for prograde flow into the left coronary artery. Left ventricular systolic function, regional wall motion and mitral regurgitation should also be assessed.

Coronary Artery Aneurysms

- Coronary artery aneurysms are one of the sequelae of Kawasaki disease.
- These can develop anywhere in the coronary arterial system. There may also be generalized dilation of the right and/or left main coronary arterial systems.
- The aneurysms may be single or multiple; size is variable, and their shape maybe fusiform, saccular or globular.
- Aneurysms may change in size, shape and distribution quite dramatically in the first month or two after the onset of Kawasaki disease.
- Large aneurysms may be filled with thrombus.

ECHO TIP:

- Sedation should be used whenever needed to improve imaging accuracy.
- *High-frequency transducers*, *minimized gain settings*, *the use of magnification* to enable a high sampling rate and accurate manipulation of the transmit focus zone are critical in evaluating the coronary arteries.
- Multiple imaging planes and windows should be used. The use of freeze-frame display and scroll-through is particularly useful.



LCA Aneurysm

Chapter 28

Connective Tissue Disorders

Connective Tissue Disorders

- This chapter illustrates cardiac involvement in systemic inflammatory disorders, such as rheumatic fever and systemic lupus erythematosos, and the cardiac manifestations of defective connective tissue formation, as with Marfan syndrome.

Marfan Syndrome

- This is a global disorder of connective tissue (fibrillin, elastin) formation with a typical pattern of cardiac involvement .
- Abnormal formation of the mitral valve apparatus and the aortic walls leads to degenerative changes that become evident with increasing age.
- The 'adult' form of Marfan syndrome typically shows gradual deterioration, with involvement of the aorta being the commonest source of morbidity.
- Progressive aortic root dilation and aneurysms, typically dissections of the thoracic aorta, are commonly seen.
- The mitral anulus maybe dilated with leaflet redundancy and scalloping.
- The subvalvular apparatus may show striking abnormalities, with elongated, redundant chordae tendineae.
- Rupture of chordae tendineae may lead to flail mitral leaflets. Aortic and mitral valve regurgitation are commonly seen.
- The 'infantile' form of Marfan syndrome, on the other hand, presents early in life. This tends to be a florid form of disease, with A-V valve (both mitral and tricuspid) regurgitation being the primary source of morbidity.
- Polyvalvular dysplasia maybe evident with infantile Marfan syndrome.

ECHO TIP:

- Best show n from parastenial long axis view.
- Each component of the mitral valve - the anulus (size), each leaflet (prolapse, redundancy, coaptation), chordae (redundancy, rupture leading to a flail leaflet), and pap illary muscles (function) - should be assessed systematically.
- The presence, timing (pansystolic, mid-systolic or late systolic) and seventy of initial regurgitation should be assessed.
- The location and width of the regurgitant jet should be defined from a parasternal short axis sweep using color flow Dopplller.
- Additional information, including left atrial and left ventricular size and the presence or absence of systolic flow reversal in the pulmonary veins - enables quantification of the severity of mitral regurgitation.
- Associated aortic root dilation should be sought using high frequency transducers from a high parasternal long axis view. *The aortic anulus, root, sinotubular junction and ascending aorta should all be measured.*

- Any linear echo dense shadows within the aortic lumen may suggest a dissecting aneurysm; these should be identified and tracked to define their extent. Localized outpouching of the aortic wall maybe the site of a true aortic aneurysm; this should be assessed with regards to size and extent.

Rheumatic Heart Disease

- Rheumatic fever may affect the heart in the acute stage, and may also leave behind catastrophic sequelae.
- Acute rheumatic c arditis may involve all of the tissue layers' of the heart; thus, it is a. pancarditis.
- Endocardial involvement maybe manifested as valvulitis leading to A-V valvar or semilunar valvar stenosis or regurgitation.
- Myocardial involvement may lead to depressed systolic function and papillary muscle dysfunction leading to A-V valve regurgitation.
- Pericardial involvement may be seen, with pericarditis and effusions.
- The longterm manifestations of rheumatic carditis most commonly involve the mitral valve. Fibrosis of the leaflets and chordae, and fusion of the papillary muscles may lead to mitral stenosis. Destruction of the mitral chordal apparatus may lead to mitral regurgitation. Occasionally, dense fibrosis of the entire valvar structure may result in a fixed-orifice mitral valve which can neither open or close completely, resulting in combined mitral stenosis and regurgitation.

ECHO TIP:

- A combination of views is needed for a conprehensive assessment.
- Every component of each A-V valve - the anulus (size), each leaflet (prolapse, redundancy, coaptation), chordae (redundancy, rupture leading to a flail leaflet), and papillary muscles (function) - should be assessed systematically.
- The presence, timing pansystolic, mid-systolic or late systolic) and severity of A-V valve regurgitation should be assessed.
- Additional information, including left atrial and left ventricular size and the presence or absence of systolic flow reversal in the systemic or pulmonary veins -enables quantification of the severity of A-V valve regurgitation.
- The aortic and pulmonary valves should also be assessed for stenosis or regurgitation.
- Left-sided valvar involvement is the commonest manifestation of rheumatic heart disease.
- While valvar involvement may be dramatic, the other components pericardium and myocardium) must also be assessed.

Systemic Lupus Erythematosus

- This is a global inflammatory connective tissue disorder that may involve pericardium, myocardium and endocardium.
- *Pericardial effusions are the commonest cardiac manifestation of lupus*. Depressed myocardial function, ventricular dilatation and mitral regurgitation may all be seen.
- The best-known form of endocardial involvement with lupus is the noninfectious Vegetation' known as the Lib man-Sacks lesion. This term refers to small vegetations on the atrioventricular valve apparatus or on the adjacent atrial or ventricular endocardium. The most characteristic location is the *ventricular surface of the posterior mitral leaflet*, where healing of the lesion may lead to adherence of the leaflet to the adjacent free wall

causing mitral regurgitation. Fibrotic thickening of the valve related to a valvulitis may also cause mitral regurgitation.

ECHO TIP:

- A complete echocardiographic assessment, including a search for pericardial effusions, assessment of valvar and ventricular function must be performed.
- If one of the cardiac manifestations of lupus is detected, additional manifestations should be sought.
- Serial evaluation is important to assess for disease progression and for the effect of medical management.

Chapter 29

Prosthetic Valves

Prosthetic Valves

- Prosthetic valves are infrequently needed in children.
- The *commonest prosthesis encountered is the St Jude va*lve, which has two semicircular leaflets that open along a hinge built into the midline of a disc.
- Other mechanical prostheses include the *ball-and-cage (Starr-Edwards)* and tilting disc and cage (Bjork-Shiley) valves.
- Porcine and other bioprostheses were used before the mechanical prostheses attained their current low-profile ease of use. Intermediate term and long term problems such as valve calcification and fibrosis have led to decreasing populairty of bioprosthetic valves.
- Mechanical Prosthetic valves pose multiple challenges for the echocardiographer. In addition to assessment of valve function, perivalvar leaks must be ruled out .
- Thrombi may attach to the valve structure, impending adequate valve function.
- Mechanical prostheses also cast metallic artefact on structures that are distal to the prothesis, making the chamber inaccessible to echocardiographic interrogation. Thus, a St Jude prothesis in the mitral position would make it very difficult to assess the left atrium from an apical view.

Normal Function

- Assessing for normal function of a prosthetic valve requires knowledge of the structure and mode of operation of the specific prosthesis.
- Thus, whether a valve is a tilting disc or a ball and cage is important information that should be known before an echocardiogram can be meaningfully interpreted.
- The flow characteristics of individual valves should also be known.

ECHO TIP:

- The best echocardiography views for visualizing a particular prosthesis depend on the location of the valve.
- Transducer rotation and angulation may be needed to clearly profile the structure and moving parts of a mechanical prosthetic valve.
- Leaflet mobility is assessed by a combination of 2-D imaging and M-mode, with the beam going through the prosthetic valve leaflets.
- Flow profiles across the prosthesis must be traced to calculate mean and peak transvalvar gradient.

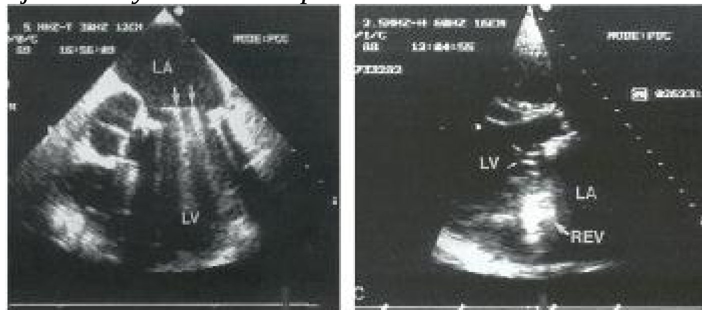


Ball-and-Cage Valves

- *Axisymmetric flow around the valve.*
- *Stagnant flow in the shadow of the ball.*



Single Disc Prosthesis
Bjork-Shiley in the Mitral position



Bileaflet Mechanical Prosthesis
St. Jude valve in the mitral position.

Obstructed Prosthetic Valve

- Prosthetic valve obstruction may be partial or complete.
- In the case of a St. Jude valve, one or both leaflets maybe immobile in the completely closed, completely open or partially openposition. This would lead to altered flow profiles across the valve.
- Knowledge of the normal flow profile across a prosthetic valve is essential to enable interpretation of abnormal flow profiles.

ECHO TIP:

- Both leaflets of the mechanical prosthesis should be profiled to enable assessment of mobility.
- Mobility of one leaflet should be compared to that of the other leaflet; any asymmetry may suggest decreased mobility of one of me leaflets.
- When both leaflets are immobile in the part-open position, the flow profile is characterized by to-and-fro flow, coupling obstructed prograde flow with the presence of regurgitant flow.

Para valvar Leak

- Paravalvar leaks are detected with increasing frequency using color flow Doppler.
- These leaks represent regurgitation around the sewing ring of the prosthetic valve; a large leak may lead to ventricular volume overload.

ECHO TIP:

- The location and diameter of the color flow jet should be defined using color flow Doppler.
- It is important to assess whether the leak originates from within the valve structure due to a thrombus that may prevent proper coaptation of the leaflets, or from around the sewing ring, constituting a true perivalvar (or paravalvar) leak.
- Careful sweeps in the short axis of the valve using color flow Doppler are essential for localizing and quantifying a perivalvar leak

Chapter 30

Post-Transplant Heart

Post-Transplant Heart

- Cardiac transplantation is increasingly Performed as 'defentive palliation for conditions ranging from end-stage cardiomyopathy to complex structural heart disease such as hypoplastic left heart syndrome.
- As the number of transplant recipients increases, there is a need for echocardiographers to identify the echocardiographic features of a normal transplanted heart.

Veins, Atria
Ventricles
Great Arteries

Veins and Atria

- The conventional method of transplantation involves leaving behind the posterior walls of the right and left atria of the recipient, and suturing the donor atria to this atrial 'cuff.
- This is termed the atrio-atrial anastomosis. It leads to the ***well-recognized biatrial enlargement*** that is one of the striking echocardiographic features of the normal transplanted heart.
- Recent concerns regarding the nonlaminar flow characteristics within the large atria post-transplant have led to the use of cavo-caval anastomosis, wherein the donor vena cavae are sutured end-to-end to the recipient vena cavae, thus minimizing the addition of atrial volume to the transplanted heart.

Echo tip:

- Best shown from apical and subcostal views.
- Obstruction at anastomotic sites may occur.
  - This should be sought specifically by careful pulsed Doppler" interrogation of the anastomotic sites. Since the vascular structure held together by cavo-caval anastomosis are narrower, they maybe more prone to obstruction than the traditional atrio atrial anastomoses.
  - Patients who have unusual systemic or pulmonary venous anatomy pre transplant (typically, patients with visceroatrial heterotaxy) may need creative and unusual vascular anastomoses as part of transplantation.

Ventricles

- The ventricles post-transplant are characterized by a hypertrophic appearance with thick walls and decreased volume. Systolic function is normal, and flattened septal motion is typical. Exaggerated translation of the entire heart with systole maybe evident, probably due to excision of the pericardium.

- Transplant Patients experiencing allograft rejection exhibit a further increase in wall thickness from baseline, and may exhibit depressed ventricular systolic function.
- Pericardial effusions may also be seen. These changes may not be specific for rejection.

Eho tip:

Best shown from parasternal long axis and apical views.

- Assessment of systolic function may be difficult due to abnormal septal motion and cardiac translation.
- Segmental wall motion abnormalities are frequently seen post-transplant. The etiology of these abnormalities is unknown.

Great Arteries

- End to end anastomoses of donor aorta and main pulmonary artery to the corresponding recipient great artery are typically performed.
- Patients with aortic arch hypoplasia (classically,patients with hypoplastic left heart syndrome) typically require extended reconstruction of the aortic arch up to and beyond the level of the ductus arteriosus. This is done by harvesting a long segment of donor aorta, and anastomosing it to the undersurface of the recipient aortic arch.

Echo tip

- The main pulmonary artery anastomosis is best shown from parasternal short axis.
- The aortic anastomosis is best shown from suprasternal views.
- Serial assessment for obstruction at all anastomotic sites is important. This is particularly important inpatients who have undergone extended aortic arch reconstruction as part of transplantation.
- Since coarctation can recur at any time after transplantation. The location and severity of recurrent coarctation must be assessed.

Chapter 31

Staged Surgery-Single Ventricle

Staged Surgery-Single Ventricle

- The goal of repair of single ventricle is completion of the Fontan operation, separating the systemic and pulmonary venous circulations.
- First Stage:
  - ü Pulmonary outflow, obstruction and decreased pulmonary blood flow may necessitate an aortopulmonary shunt.
  - ü A hypoplastic aortic arch necessitates arch reconstruction (the Norwood operation) as the first stage of repair.
  - ü Subvalvar aortic obstruction without arch hypoplasia (e.g., due to a restrictive bulboventricular foramen, as discussed in Chapter 20) may necessitate the Damous-Kaye-Stansel procedure to bypass the obstruction.
  - ü Patients who have neither aortic nor pulmonary outflow obstruction may need a pulmonary artery band to control pulmonary flow.
- Subsequent stages culminate in complete separation of the systemic and pulmonary venous circulations. Most patients with a functional single ventricle would undergo a bi-directional Glenn shunt as an intermediate stage prior to completion of the Fontan operation.

Pulmonary Artery Band
Aortopulmonary shunt
Norwood Stage I
Damus-Kaye-Stansel Procedure
Bi-directional Glenn
Fontan

Pulmonary Artery Band

Ø CLASSICAL INDICATIONS

§ Originally, pulmonary artery banding was suggested for patients with ''functional single ventricle'', i.e. those with a large VSD or a single ventricle. With the development of open-heart surgery, symptomatic VSDs were more routinely closed during intracardiac operations.

§ However, as the risk of VSD closure in sick infants with other associated defects remained high, pulmonary artery banding was commonly used as a palliative procedure to reduce pulmonary arterial pressure and blood flow in this age group. Nevertheless, the disadvantages of too tight a band and the potential distortion of the pulmonary bifurcation and the valve were well appreciated.

§ Other conditions *causing increased pulmonary blood flow* have also been palliated by banding:

- ü acyanotic lesions (atrio-ventricular septal defect, double-inlet ventricle,

- ü and some forms of double-outlet ventricle without pulmonary stenosis)
- ü and cyanotic lesions, including transposition of the great arteries (TGA) with VSD,
- ü some forms of tricuspid atresia, truncus arteriosus, congenitally corrected transposition with VSD, hypoplastic left heart syndrome and mitral atresia.
- ü Since early primary repair of VSD, TGA and several other lesions became the treatment of choice, pulmonary artery banding has been used less frequently. However, the latter trend was not immune to changes.

Ø CURRENT INDICATIONS:

- § Ventricular Septal Defect Most VSDs are submitted to rigorous medical treatment in the first instance of severe heart failure.
  - ü If medical treatment fails, primary closure of such defects .
  - ü There are, however, a few exceptions in which PAB is considered:

1. *Multiple VSDs* in early infancy. Surgery remains risky, although new approaches, such as doublepatch sandwiching the septum (Brizard *et al*., 2004), have been reported, with encouraging results.
2. *Coarctation of the aorta with a large VSD*. In these patients, coarctation of the aorta is repaired.
3. *VSD complicated by other congenital non-cardiac anomalies* (e.g. large omphalocoele, oesophageal atresia, diaphragmatic hernia, syndromal anomalies, premature baby with low birth weight, or VSD in severely undernourished babies and the presence of severe infection (sepsis, respiratory, skin) that cannot be brought under control because of severe heart failure).

- § Transposition of the Great Arteries
- ü *The indications for PAB for patients with TGA have changed considerably*. At present, there is no strong indication for PABin infants withTGAand VSD, except for occasional patients with multiple VSDs and additional severe noncardiac malformations.
- § Double-inlet Ventricle and Tricuspid Atresia without Pulmonary Stenosis In the absence of pulmonary stenosis, patients with double-inlet ventricle and tricuspid atresia present in heart failure. A Fontan-type procedure or total cavopulmonary connection (TCPC), is usually the only definitive solution. Staged approaches with a bidirectional superior cavo-pulmonary anastomosis (Glenn shunt) or a hemi-Fontan are currently not favoured for patients under the age of 3 months because the pulmonary vascular resistance is still too high. If the pulmonary blood flow is not restricted and the pulmonary arterial pressure not reduced, pulmonary resistance continues to increase. PAB is therefore indicated.
- § Other Indications PAB for double-outlet right ventricle, truncus arteriosus, and complete atrio-ventricular septal defect has been largely *abandoned* in favour of early complete repair. Under special circumstances (such as those outlined for VSD, late-presenting atrio-ventricular septal defect with pulmonary hypertension, straddling atrio-ventricular valves, and atrio-ventricular discordance), PAB may be considered.
- § Hypoplastic Left Heart (HLH) The surgical concept, with bilateral PAB, preceded by endovascular stenting of the patent ductus arteriosus, is being very selectively applied as part of the Norwood strategy in the management of HLH. This new approach, together with atrial septectomy or septostomy, was first reported by Gibbs *et al*. (1993). It has been used in very sick neonates with HLH to improve the haemodynamics before a delayed stage 1 Norwood procedure (Pizarro and Norwood, 2003).

ECHO TIP:

- Best shown from parasternal short axis and subcostal views.
- Band location must be defined, including its proximity to the pulmonary valve leaflets and to the branch pulmonary arteries.
- Band encroachment onto the origin or the lumen of either branch pulmonary artery, and any erosion of die band into the pulmonary arteries must be identified.
- Serial assessment of band location is important since migration of the band either proximally or distally can occur with linear growth.
- Peak flow velocity across the band must be recorded together with the systolic blood pressure.
- In a patient with single ventricle and no aortic outflow obstruction, these measurements enable estimation of systolic pressure in the pulmonary artery distal to the band. In a patient with two ventricles and a ventricular septal defect who is being staged towards the Fontan operation (as with double outlet right ventricle and a canal-type ventricular septal defect), the peak velocity of flow across the ventricular septal defect must also be measured to allow estimation of pulmonary artery pressure.

Aortopulmonary Shunts

- This procedure is needed for patients who have a *functionally single ventricle* with decreased pulmonary blood flow.
- The purpose of this operation is to provide volume - and pressure - controlled augmentation of pulmonary blood flow.
- The shunt typically consists of a precisely sized Goretex tube extending from the side of the subclavian artery to the side of ipsilateral branch pulmonary artery. This is termed a *modified Blalock-Taussing shunt*; the classic Blalock-Taussig shunt was a direct connection of the end of the (transected) subclavian artery to the side of the ipsilateral branch pulmonary artery.
- Other shunts that are used currently include a central shunt, which connects the ascending aorta to the main pulmonary artery, and a variation of the Blalock-Taussig shunt connecting the innominate artery to the right pulmonary artery.

ECHO TIP:

- Best shown from suprasternal short axis and angled suprasternal long axis views.
- The entire length of the shunt should be visualized, since shunt obstruction or stenosis may occur anywhere along the length of the shunt.
- Shunt diameter should be measured at several points, particularly where stenosis is suggested by imaging or color Doppler.
- Distortion or stenosis of the branch pulmonary arteries should be sought, particularly in the vicinity of shunt insertion into the pulmonary arteries.
- Peak flow velocity in the shunt should be measured, with concomitant recording of systolic blood pressure. These measurements enable estimation of pulmonary arterial systolic pressure.
- The caveat is that this technique probably underestimates the gradient (and overestimates pulmonary artery pressure) due to the long tubular nature of the shunt.

Norwood Stage I

- The goals of this operation are to allow the *right ventricle to pump unobstructed into the systemic circulation*, to establish unrestricted mixing at the atrial level, and to provide volume - and pressure - controlled pulmonary blood flow.
- This operation consists of aortic arch augmentation using the main pulmonary artery usually in a side-to-side anastomosis. The main pulmonary artery is divided, thus eliminating prograde flow into the pulmonary arteries.
- An aortopulmonary shunt is placed, usually from the innominate artery to the right pulmonary artery.
- An atrial septectomy is performed to ensure adequate mixing at the atrial level.

ECHO TIP:

- Post-operative echocardiograms should assess for pulmonary artery growth, distortion and stenosis.
- Pulmonary arterial pressures should be assessed using the peak systolic gradient across the shunt, keeping in mind that the gradient is probably an underestimate due to the length of the shunt.
- The neoaortic arch should be interrogated for coarctation in its entirety into the descending aorta, since coarctation can occur at any location along the anastomosis.
- The adequacy of the interatrial communication must be assessed.
- Systemic (right) ventricular function and the presence and severity of systemic A-V (tricuspid) valve regurgitation must also be assessed.
- Patency of the entire length of the aortopulmonary shunt must also be confirmed.

Dainus - Kaye - Stansel Procedure

- This procedure is needed for patients who have a *functionally single ventricle with subaortic obstruction* and a well-developed aortic arch.
- This typically occurs with a single left ventricle and absent right ventricle, where the pulmonary artery arises from the single ventricle and the aorta arises from the infundibular chamber.
- The communication between the single ventricle and the infundibulum, the bulboventricular foramen, maybe restrictive, thus causing aortic outflow tract obstruction.
- The Damus-Kaye-Stansel procedure uses the pulmonary outflow tract and main pulmonary artery to bypass this obstruction.
- Thus, the main pulmonary artery is transected; its distal end is oversewn, and the cardiac end is anastomosed to the side of the ascending aorta; prosthetic material maybe needed to augment this anastomosis. Pulmonary blood flow is provided via an aortopulmonary shunt .Following this operation, both semilunar valves (the native aortic valve and the neo-aortic (native pulmonary) valve function as "aortic" valves.

ECHO TIP:

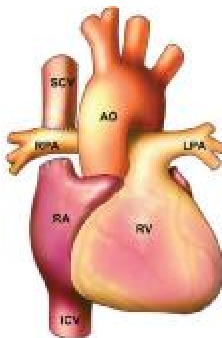
- Best shown by a combination of parasternal short axis and sub costal views.
- Visualizing these anastomoses can be a challenge due to the three-dimensional nature of the bypassing conduit.
- Conduit diameter should be measured and obstruction to flow within the conduit should be sought.
- Since both semilunar valves now function as 'aortic'valves, any regurgitation of either valve represents aortic regurgitation and should be assessed carefully. The native pulmonary (neo-aortr) valve is particularly prone to distortion and regurgitation.

Bidirectional Glenn Shunt

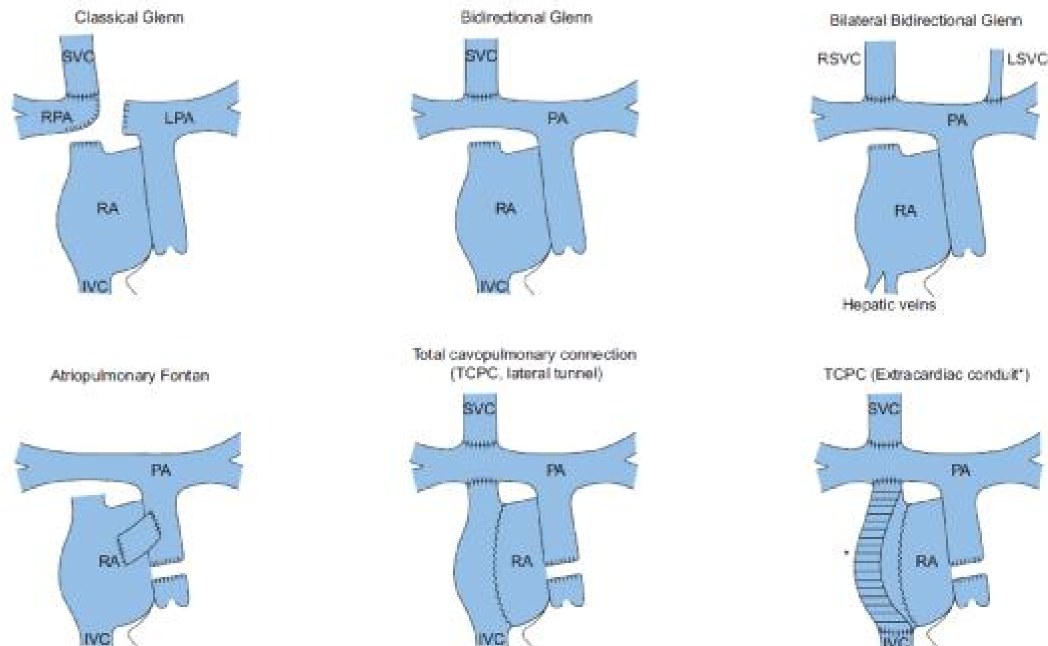
- The goals of this operation are to provide an intermediate stage to the Fontan operation, while decreasing systemic ventricular volume loading.
- The components of this operation include division of any aortopulmonary shunt that may have been placed.
- The pulmonary arteries are isolated if this has not already been done.
- The bidirectional Glenn shunt is created by suturing the distal end of the divided superior vena cava to the side of the right pulmonary artery, and oversewing the cardiac end of the superior vena cava. Additional surgery is performed as needed to correct atrial septal restriction, pulmonary arterial distortion, atrioventricular valve regurgitation or aortic arch obstruction.
- The specifics of the operation may differ based on systemic venous anatomy in the individual patient. Thus, if bilateral superior vena cavae exist, bilateral bidirectional Glenn shunts maybe needed, in some situations with a small but usable pulmonary ventricle, prograde flow from the pulmonary ventricle into the pulmonary arteries may be maintained (the so-called powered Glenn shunt).

ECHO TIP:

- Echocardiograms obtained prior to the Glenn shunt should assess for pulmonary artery growth, distortion and stenosis.
- Inpatients who have had aortic arch reconstruction, the entire neoaortic arch should be interrogated for coarctation which may occur at any location along the anastomosis.
- The adequacy of the interatrial commmucation, systemic ventricular size and function and the presence and severity of systemic A-V and aortic valve regurgitation must also be assessed. Patency of the entire length of the aoriopulmonary shunt, must also be confirmed.
- Pulmonary arterial pressures should be assessed using the peak systolic gradient across the shunt, keeping in mind that the gradient is probably an underestimate due to the length of the shunt.
- Echocardiograms obtained after the Stage 2 procedure should, in addition to the above, assess for patency of the Glenn shunt *This shunt is best shown from a suprasternal short axis view, angling rightwards*.
- Since the systemic veins have high compliance, obstruction of a bidirectional Glenn shunt maybe more evident on the *basis of dilation of the superior vena cava and azygous vein* (if patent) than by Doppler gradients.
- It is difficult if not impossible to assess pulmonary artery pressure by echocardiography following a Glenn shunt; *pulmonary hypertension is suggested by a dilated superior vena cava with no evidenc of obstruction*.



Bidirectional cavo-pulmonary anastomosis, bidirectional Glenn procedure. SCV, Superior caval vein; IVC, inferior caval vein;RPA, right pulmonary artery; LPA, left pulmonary artery;AO, aorta; RA, right atrium; RV, right ventricle.



*Types of venous anastomoses and Fontan operations. See text. RPA, right pulmonary artery; SVC, superior vena cava; LPA, left pulmonary artery;RA, right atrium; IVC, inferior vena cava; PA, pulmonary artery; RSVC, right superior vena cava; LSVC, left superior vena cava; TCPC, total cavo-pulmonary connection; *, conduit or prosthetic tube.*

Fontan Operation

- This is the final stage in the surgical palliation for patients with functional single ventricle.
- The goals of surgery are to achieve complete separation of the pulmonary and systemic circulations.
- This operation consists of placement of a baffle or tunnel that directs hepatic venous and inferior vena caval blood into the right pulmonary artery.
- Thus, the combination of this operation with the bidirectional Glenn shunt would channel superior and inferior vena caval return as well as hepatic venous return into the pulmonary arteries directly.
- Additional surgery is performed as needed to correct atrial septal restriction, distortion or stenosis of the Glenn shunt or of the pulmonary arteries, aortic arch obstruction, or neoaortic or tricuspid valve regurgitation.
-

ECHO TIP:

- Pre-Fontan echocardiograms should *assess* for pulmonary artery growth, distortion and stenosis.
- Patients who have previously undergone aortic arch reconstruction should have a complete assessment of the neo aortic arch for coarctation.
- The adequacy of interatrial communication, systemic ventricular size and function, and the presence and severity of systemic A-V and aortic valve regurgitation must also be assessed.

- Patency of the entire length of the Glenn shunt should be confirmed; obstruction of this low-pressure system may be more evident on the basis of 2-D imaging of the obstruction and a dilated superior vena cava and azygous vein (if patent) than on the basis of Doppler gradients. *A dilated superior vena cava with no evidence of obstruction should suggest pulmonary hypertensio*n.
- Post-Fontan echocardiograms should, in addition to the above, also assess for patency of the lateral tunnel. *This is frequently difficult to visualize; it is best shown by a combination of subcostal and suprasternal imaging*. Since the systemic veins have high compliance, *obstruction of a Fontan baffle may be more evident on the basis of dilated hepatic veins and inferior vena cava than on the basis of Doppler gradients*.
- Again, dilated systemic veins with no evidence of obstruction should suggest pulmonary hypertension.

Chapter 32

Switch Procedures

Switch Procedures

- The purpose of a switch operation, whether at the atrial or great arterial level, *is to undo transposition physiology*.
- The lesion that has been the prototype for such operations is {S,D,D} transposition of the great arteries.
- The atrial switch (Mustard and senning) procedures were employed previously; at the present time, the arterial switch operation is the procedure of choice unless unusual mitigating circumstances occur. This chapter illustrates the principles and anatomy of the various switch operations.

Arterial Switch

- The arterial switch operation entails transection (anatomical correction)of both great arteries followed by reanastomosis of each distal great artery to the opposite-sided root; thus, the distal aorta is anastomosed to the great arterial root originating from the left ventricle (the native pulmonary root).
- The coronary arteries are mobilized with a cuff of aortic wall, and are anastomosed to this neo-aorta.
- Since most but not all cases of d-transposition are associated with an anterior aorta and a posterior pulmonary artery, the distal main pulmonary artery would be too short to be connected to the anteriorly placed right ventricular outflow tract for completion of the arterial switch. To enable this connection, the pulmonary arterial confluence is brought anterior to the ascending aorta (the Lecompte maneuver).

ECHO TIP:

- Best shown from subcostal long axis and parasternal short axis sweeps.
- The branch pulmonary arteries have an unusual origin and course in this condition.
- The pulmonary arterial confluence is anterior to the ascending aorta, and the proximal branch pulmonary arteries wrap around the ascending aorta as they dive posteriorly.
- Early postoperative issues for the echocardiographer include assessment of left ventricular function. This maybe compromised due to coronary artery obstruction or kinking.
- Other causes of early postoperative left ventricular dysfunction may include an inadequately pressure-primed left ventricle in an older (> 3-4 weeks old) infant.
- Supravalvar pulmonary stenosis or, less commonly, supravalvar aortic stenosis at the anastomotic sites in the great arteries, should be assessed.
- Branch pulmonary artery stenosis or diffuse hypoplasia can occur, particularly involving the left pulmonary artery, which is stretched antero-posteriorly with the Lecompte maneuver.

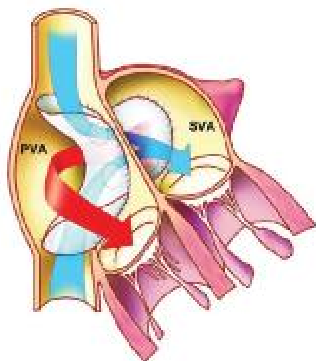
- Neo-aortic valve insufficiency, neo-aortic root dilation, growth of the coronary anastomosis and left ventricular function are other long-term issues which emphasize the need for serial, complete echocardiographic follow up.

Atrial Switch -Mustard

- The Mustard and Semung operations work by 'switching' the ventricular inflows.
- Both these operations utilize baffles that channel superior and inferior vena caval blood to the mitral valve, the left ventricle and the pulmonary artery. The baffles also direct pulmonary venous drainage to the tricuspid valve, the rigid ventricle and the aorta.
- The 'atrial switch' therefore results in a physiologic correction, in that the pulmonary and systemic circulations are placed in series. However, the tricuspid valve and right ventricle are subjected to systemic loading conditions, which can result in longterm problems with tricuspid regurgitation and right ventricular dysfunction.
- The Mustard and Senning operations differ in the specifics of baffle construction. The *Mustard operation utilizes an atrial septectomy*; the baffles are constructed from autologous pericardium or prosthetic material. In the *Senning procedure, tissue flaps from the right atrial wall and atrial septum are used to construct the baffles*.

ECHO TIP:

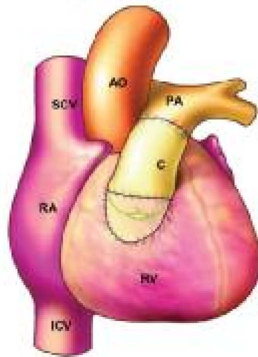
- Echocardiography *cannot reliably determine* the difference between (the Mustard and Senning procedures. The importance of knowing which procedure has been performed on a given patient lies in their differing spectra of longterm complications.
- The *superior vena cava baffle is best shown from the apical view* with clockwise rotation of the transducer. The *inferior vena cavalbaffle is best shown from parasternal short axis view*, running in the posterior wall of the right atrium. In smaller children, the subcostal long axis view with counterclockwise rotation of the transducer is an additional imaging window. The pulmonary venous baffle is best shown from the apical view and subcostal long axis view.
- Baffle obstruction is commoner after the Mustard *operation*, particularly involving the superior vena caval baffle where it makes an acute turn past the atrial septum.
- Inferior vena caval baffle obstruction is rare, and usually occurs between the mouth of the coronary sinus and the right tower pulmonary vein. Pulmonary venous obstruction usually occurs anterior to the entrance of the right pulmonary veins.
- Residual baffle leaks may occur anywhere along the suture lines, resulting in a shunt in either direction.
- Right ventricular hypertrophy is the rule after an atrial switch operation. Right ventricular dysfunction and tricuspid regurgitation are longterm complications that require lifelong surveillance.



Mustard correction of transposition of the great arteries. Blue arrow shows the venous flow from superior and inferior caval veins through intra-atrial baffles to systemic venous atrium (SVA)and mitral valve orifice. Red arrow shows pulmonary venous flow redirected into pulmonary venous atrium (PVA) to tricuspid valve.

Rastelli operation

- Patients with TGA, VSD, and severe fixed valvular or subvalvular pulmonary stenosis may undergo anatomical correction according to Rastelli .
- This operation has the advantage of incorporating the left ventricle as the systemic ventricle.
- A large intraventricular baffle is sutured into place, closing the VSD and redirecting the left ventricular outflow to the more anterior placed aortic valve.
- The connection between the left ventricle and the pulmonary artery is closed and a valved conduit (a valved Hancock Dacron conduit, a porcine xenograft or a human homograft) is placed between the right ventricle and the pulmonary artery .



Rastelli correction of transposition of the great arteries.
ICV, Inferior caval vein; SCV, superior caval vein; RA, right atrium;RV, right ventricle; AO, aorta; PA, pulmonary artery;LA, left atrium; LV, left ventricle; C, conduit from right ventricle to pulmonary artery.



Transposition of the great arteries, Rastelli correction performed in adulthood; transthoracic echo. Conduit (C) with bioprothesis from the right ventricle to pulmonary trunk-bifurcation.

Chapter 33

Pulmonary Hypertension

Pulmonary Hypertension

- When measured directly in the cardiac catheterization laboratory, the normal pulmonary artery (PA) systolic pressure of children and adults is _30 mm Hg and the mean PA pressure is _25 mm Hg at sea level. A diagnosis of pulmonary hypertension can be made when the mean PA pressure is >25 mm Hg in a resting individual at sea level. The PA pressure is higher at high elevations.
- The noninvasive Doppler method, however, often overestimates the PA pressure and may even suggest PA hypertension in people who are normal.
- Using tricuspid regurgitation jet velocity and the modified Bernoulli equation, with right atrial pressure assumed to be 10 mm Hg, the mean PA systolic pressure (±SD) was found to be 28.3 ± 4.9 mm Hg (range 15 to 57 mm Hg) in infants and adults, higher values than previously reported using invasive methods (McQuillan et al, 2001). The estimated upper 95% limit for PA systolic pressure was 37.2 mm Hg. (This results from a tricuspid regurgitation jet velocity of 2.7 m/second in the absence of pulmonary stenosis.) *Thus, a Doppler-estimated PA systolic pressure of 36 to 40 mm Hg has been assumed as a cutoff value for mild PA hypertension.*

CAUSES OF PULMONARY HYPERTENSION

- The causes of pulmonary hypertension can be grouped into the following five.

1. Increased pulmonary blood flow (PBF) seen in congenital heart diseases (CHDs) with large left-toright shunts (hyperkinetic pulmonary hypertension)
2. Alveolar hypoxia
3. Increased pulmonary venous pressure
4. Primary pulmonary vascular disease
5. Other diseases that involve pulmonary parenchyma or pulmonary vasculature

Hyperkinetic Pulmonary Hypertension

- Pulmonary hypertension associated with large left-to-right shunt lesions, such as ventricular septal defect (VSD) and patent ductus arteriosus (PDA), is called *hyperkinetic pulmonary hypertension*.
- It is the result of an increase in pulmonary blood flow, a direct transmission of the systemic pressure to the PA, and an increase in pulmonary vascular resistance by compensatory pulmonary vasoconstriction. If no vasoconstriction occurs, the increase in pulmonary blood flow is much larger and an intractable congestive heart failure (CHF) results.
- Defects in the vasodilation machinery of the endothelial cell, such as overproduction of vasoconstrictor elements, have been implicated in this form of pulmonary hypertension.
- Hyperkinetic pulmonary hypertension is usually reversible if the cause is eliminated before permanent changes occur in the pulmonary arterioles (see later section).
- If large left-to-right shunt lesions (such as VSD, PDA, complete atrioventricular canal) are left untreated,irreversible changes take place in the pulmonary vascular bed, with severe

pulmonary hypertension and cyanosis because of a reversal of the left-to-right shunt. *This stage is called Eisenmenger's syndrome or pulmonary vascular obstructive disease (PVOD)*. Surgical correction is not possible at this stage.

- The time of onset of PVOD varies, ranging from infancy to adulthood, but the majority of patients develop PVOD during late childhood or early adolescence. It develops even later in patients with atrial septal defect. Many patients with transposition of the great arteries begin to develop PVOD within the first year of life for reasons not entirely clear. Children with Down syndrome with large left-to-right shunt lesions tend to develop PVOD much earlier than normal children with similar lesions.

Pulmonary Venous Hypertension

- Increased pressure in the pulmonary veins produces reflex vasoconstriction of the pulmonary arterioles,raising PA pressure to maintain a high enough pressure gradient between the PA and the pulmonary vein.
- This pressure gradient maintains a constant forward flow in the pulmonary circulation.
- There is a marked individual variation in the degree of reactive pulmonary arteriolar vasoconstriction. For example, when the pulmonary venous pressure is elevated in excess of 25 mm Hg from MS, marked reactive pulmonary hypertension occurs in less than one third of patients.
- The mechanism for the vasoconstriction is not entirely clear, but a neuronal component may be present. Moreover, an elevated pulmonary venous pressure may also narrow or close small airways, resulting in alveolar hypoxia, which may contribute to the vasoconstriction.
- MS, total anomalous pulmonary venous return (TAPVR) with obstruction (of pulmonary venous return to the LA), and chronic left-sided heart failure are examples of this entity.
- Pulmonary hypertension with increased pulmonary venous pressure is usually reversible when the cause is eliminated,with the exception of congenital pulmonary vein stenosis, for which no curative surgery is available.

ECHO TIP:

- Echo usually demonstrates the following.
    1. Enlargement of the RA and RV, with normal or small LV dimensions.
    2. Thickened interventricular septum and abnormal septal motion (as a result of the RV pressureoverload).
    3. Thickened RV free wall and RV dysfunction are difficult to demonstrate and quantitate.
- *Semiquantitative estimation of PA pressures* can be obtained using various methods such as M-mode or two dimensional echo and Doppler examination. It should be noted, however, that the Doppler-estimated PA pressure and that measured in the cardiac catheterization laboratory are not interchangeable (as discussed earlier in this chapter).
- These noninvasive methods of estimating the severity of pulmonary hypertension are presented in the following.

1.*M-Mode*.

- *Abnormal valve motion on M-mode echo*:
- An absent or diminished *a* wave, a reduced EF slope, and
- a midsystolic closure (notching) indicate pulmonary hypertension .
- However, these abnormalities are not always present, and a false-positive result occurs rarely.

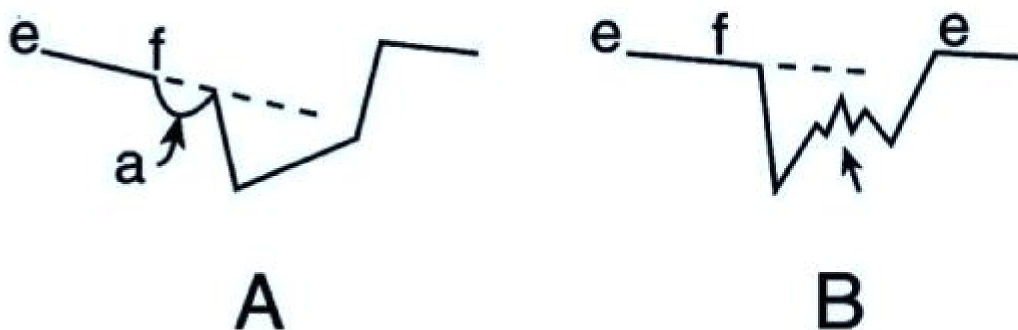
2. *Two-dimensional echo*: With an elevated RV pressure, the interventricular septum shifts toward the LV and appears flattened at the end of systole. An inspection of the septal curvature at the end of systole provides an estimate of the RV systolic pressure .

3. *Doppler echo*:

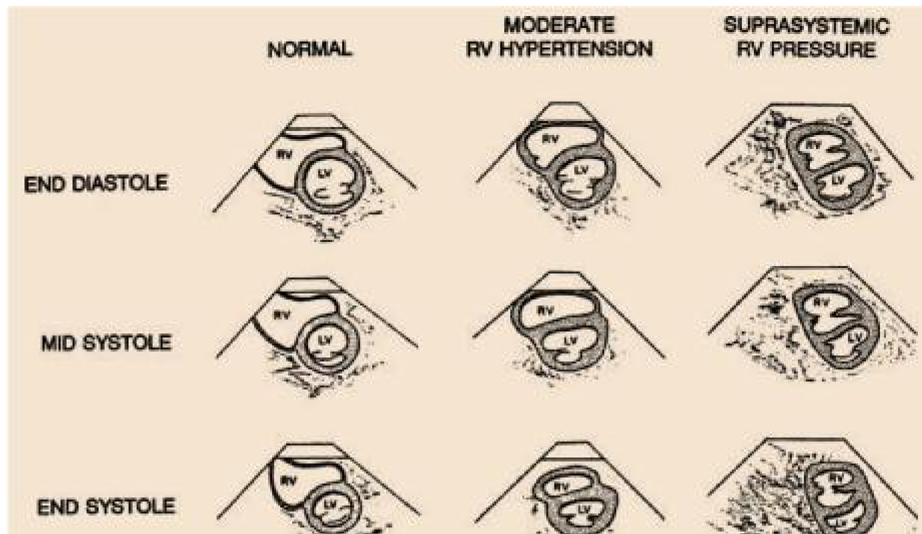
a. *Peak TR velocity* determined by continuous-wave Doppler is used to estimate the *systolic* pressure in the PA. The simplified Bernoulli equation (_P = 4V2) is used to estimate a systolic pressure drop across the tricuspid valve; a normal central venous pressure of 10 mm Hg is added to the result to estimate PA *systolic* pressure. (Note that the normal Doppler-derived values are different from those obtained by invasive methods; the upper limit of normal PA systolic pressure is 36 to 40 mm Hg by Doppler method.) In the absence of pulmonary stenosis, the systolic pressure in the RV equals that in the PA.

b. *With a shunt lesion*, such as VSD, PDA, or systemic-to-PA shunt, the peak systolic velocity across the shunt can be used to estimate systolic pressure in the RV or PA. The systolic pressure in the LV (which is equal to the aortic pressure) estimated by systolic pressure in the arm, minus the systolic pressure drop across the VSD or PDA, estimates RV and PA systolic pressures, respectively. Note that the systolic pressure in the arm is a little higher (5 to 10 mm Hg) than the LV systolic pressure because of the peripheral amplification of systolic pressure .

c. The *end-diastolic velocity of PR* can be used to estimate the *diastolic* pressure in the PA. The end-diastolic (not early diastolic) velocity is measured and entered into the modified Bernoulli equation, and a normal central venous pressure of 10 mm Hg is added.



M-mode echo of the pulmonary valve in pulmonary hypertension. A, Normal M-mode echo. B, Pulmonary hypertension demonstrating an absent a wave, diminished or negative EF slope, and midsystolic notch or flutter (arrow).



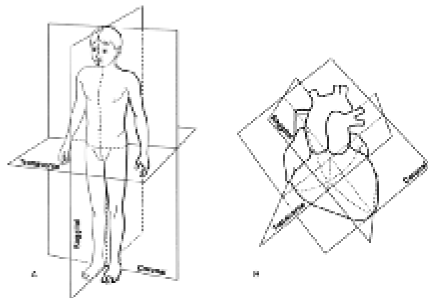
Parasternal short-axis stop frames of interventricular septal configurations of normal patients and patients with right ventricular (RV) hypertension. The top row represents the end-diastolic frames, the middle row represents the midsystolic frames, and the bottom row represents the end-systolic frames. In normal children (left column), the typical rounded configuration of the interventricular septum is demonstrated throughout the cardiac cycle. In moderate pulmonary hypertension (middle column), the interventricular septum becomes progressively flattened from the end of diastole to the end of systole.

When RV pressure is suprasystemic (right column), the interventricular septum is flattened at the end of diastole; at the end of systole, it reverses its curvature to become convex toward the left ventricle. *(Modified from King ME, Braun H, Goldblatt A, et al: Interventricular septal configuration as a predictor of right ventricular systolic hypertension in children: A cross-sectional echocardiographic study. Circulation 68:68-75, 1983.)*

Appendex1

Planes of the Heart and Technique of Sweeping

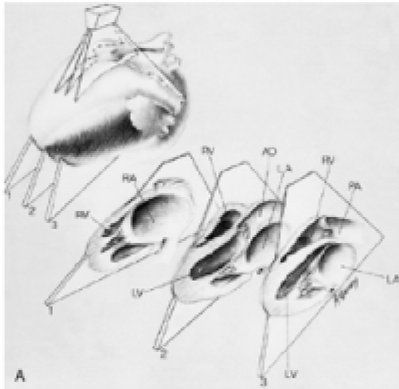
- Planes of the Heart and Technique of Sweeping: Thinking in Three Dimensions Three-dimensional (3-D) imaging continues to improve (see below) but is currently not in routine clinical use.
- Until it is, the challenge and essence of pediatric echocardiography are and will be acquiring all the necessary 2-D images, ***mentally synthesizing them into a 3-D model***, and conveying this 3-D representation to others by narrative or visual tools.
- The spatial location of any part of an object is defined and understood by considering it in relation to the three planes (transverse [axial], sagittal, and coronal) in which the object exists.
- Each of the four echocardiographic windows affords the opportunity to image the heart from one or more of these three planes.
- From the parasternal window, the long (sagittal) and short (axial) planes are shown.
- From the apical and subcostal windows, the four-chamber (coronal) and two-chamber (sagittal) planes are demonstrated.
- Finally, from the suprasternal notch window, the sagittal and axial axes of the upper thoracic vasculature are imaged.
- Sweeping the transducer through the nearly parallel planes within each of the acoustic windows mimics the ability of other imaging modalities such as magnetic resonance to obtain parallel slices within a given plane . With these techniques, the spatial relationships become clear and the 3-D mental reconstruction of the heart becomes possible.



There are three imaging planes of the body: sagittal, coronal, and transverse. B: There are also three imaging planes of the heart. The cardiac imaging planes are rotated leftward and anterior because the heart's axes are rotated leftward and anterior relative to the body's.

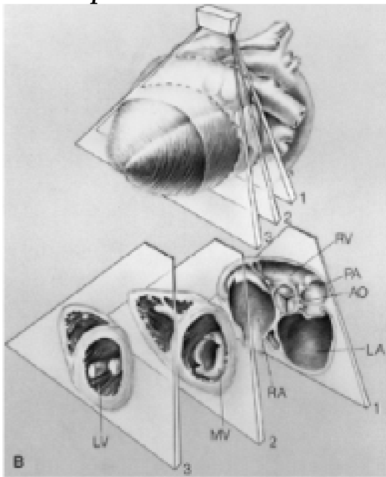
The family of sweeps for each of the imaging planes of the heart

A: *The parasternal long-axis sweeps*:



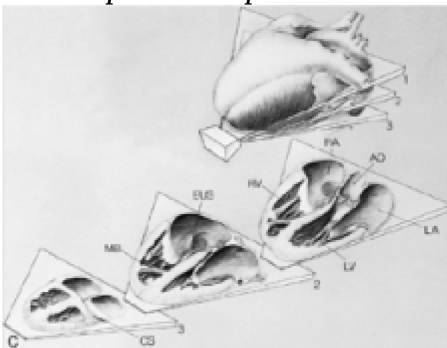
consist of the rightward tricuspid valve view (1), the standard long-axis plane (2), and the leftward pulmonary valve view (3).

B: *The parasternal short-axis sweeps*



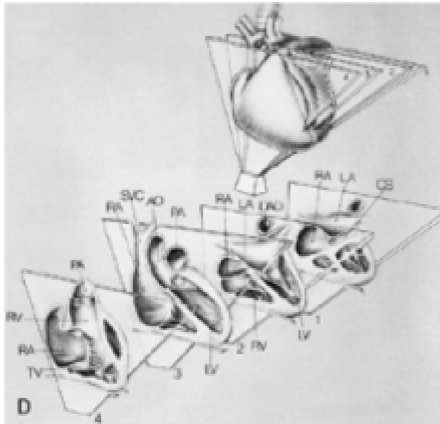
consist of the superior basal view (1), the standard plane at the level of the mitral valve (2), and the inferior papillary muscle view (3).

C: *The apical sweeps*:



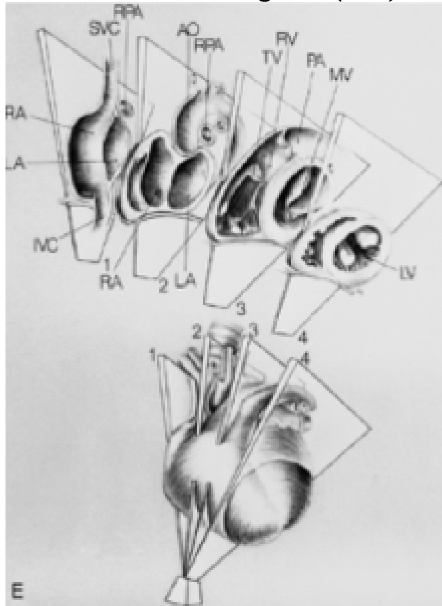
consist of the anterior five-chamber view (1), the standard apical four-chamber view (2), and the posterior coronary sinus view (3).

D: *The subcostal coronal (long axis)sweeps*:3:00 clock



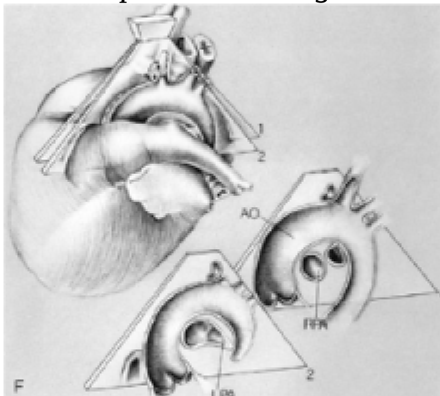
consist of the posterior coronary sinus view (1), the standard four-chamber view (2), the anterior left ventricular outflow tract view (3), and the extremely anterior right ventricular outflow tract view (4).

E: *The subcostal sagittal(sax) sweeps* :6:00 clock



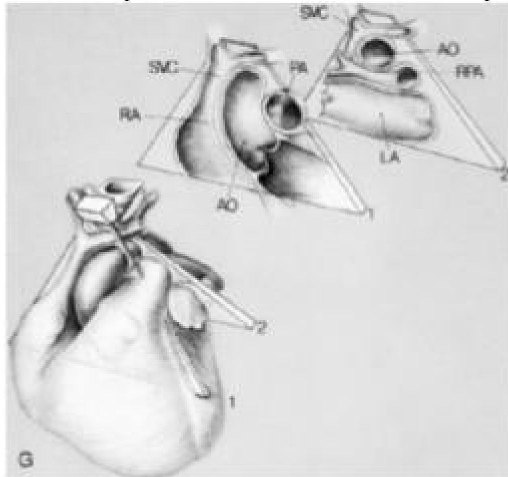
consist of the rightward systemic venous return view (1), the slightly leftward left ventricular outflow tract view (2), the leftward right ventricular outflow tract view (3), and the extremely leftward ventricular view (4).

F: *The suprasternal long-axis sweeps*:12:00 clock



consist of the standard aortic arch view (1), the rightward superior vena caval view (not shown), and the leftward left pulmonary artery view (2).

G: *The suprasternal short-axis sweeps*:3:00 clock



consist of the very anterosuperior strap vessels view (not shown), the anterosuperior vena cava and innominate vein view (1), the standard right pulmonary artery and left atrial view (2), and the posterior descending aorta view (not shown).

AO, aorta; CS, coronary sinus; DAO, descending aorta; EUS, eustachian valve; LA, left atrium; LPA, left pulmonary artery; LV, left ventricle; MB, muscle bundle; MV, mitral valve; PA, pulmonary artery; RA, right atrium; RPA, right pulmonary artery; RV, right ventricle; SVC, superior vena cava; TV, tricuspid valve.

Pediatric views

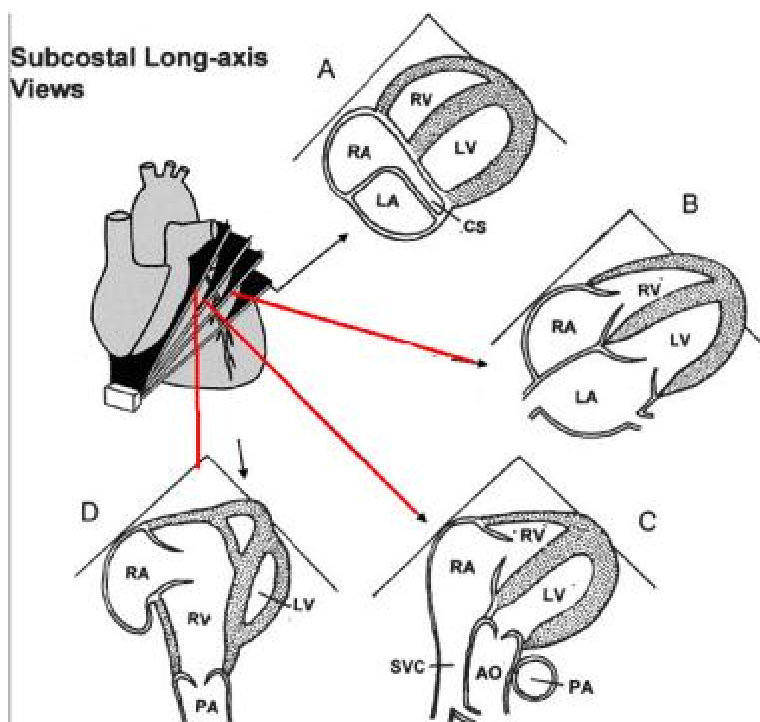


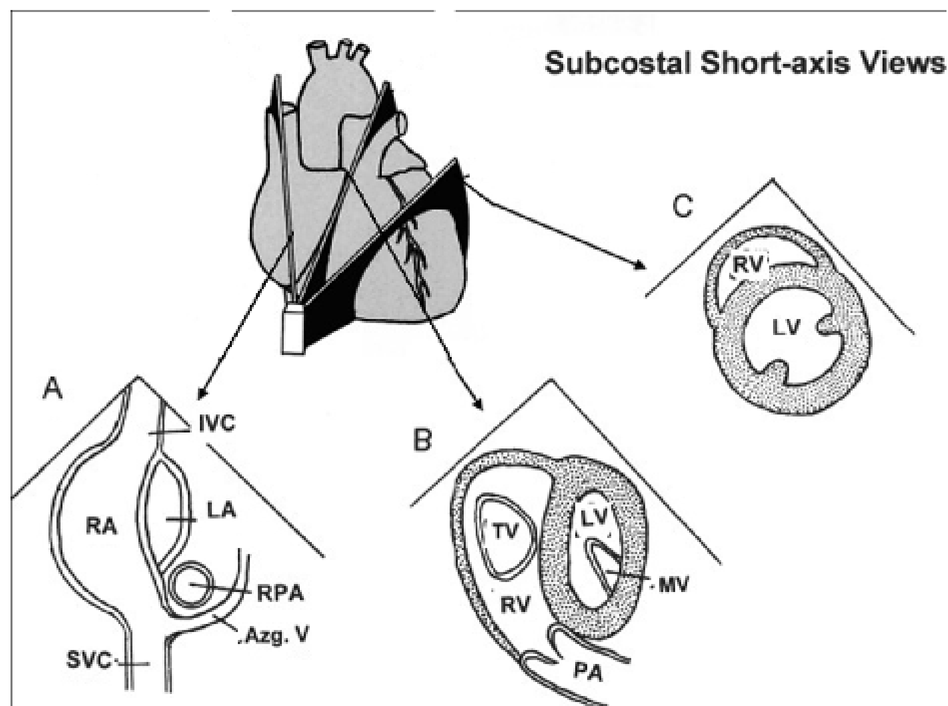
Diagram of subcostal long-axis(coronal) views.
A, Coronary sinus view posteriorly.

Subxiphoid Long Axis View Coronary Sinus Plane
* *TRANSDUCER placed just below xiphoid.*
* *INDICATOR is at 3:00.*
* *Posterior tilt to image coronarv sinus.*

B, Standard subcostal four-chamber view.
C, View showing the left ventricular outflow tract and the proximal aorta.
Subxiphoid Long Axis View LVOT and Ao plane
- *Transducer located in the abdomen just below xyphoid process*
- *Indicator is at 3:00*
- *Slight anterior tilt to image LVOT and Ao*

D, View showing the right ventricular outflow tract (RVOT) and the proximal main pulmonary artery.
Sabxiphoid Long Axis View
RVOTandPA Plane
- *Transducer located in the abdomen just below xyphoid process*
- *Indicator is at 3:00*
- *Slight anterior tilt to image ROVT and PA*

AO, aorta; CS, coronary sinus; LA, left atrium; LV left ventricle; PA,pulmonary artery; RA, right atrium; RV right ventricle, SVS, superior vena cava.



Subcostal short-axis (sagittal) views.
A, Entry of venae cavae with drainage of the azygos vein.
Subxiphoid Short Axis View
Caval Plane(SVC IVC Plane) Transducer located in the abdomen just below xephoid process.
Rotated clockwise from long axis(opposite adult).Indicator is at 6:00.
Tilt US beam towards patient's right shoulder. Put a little twist.

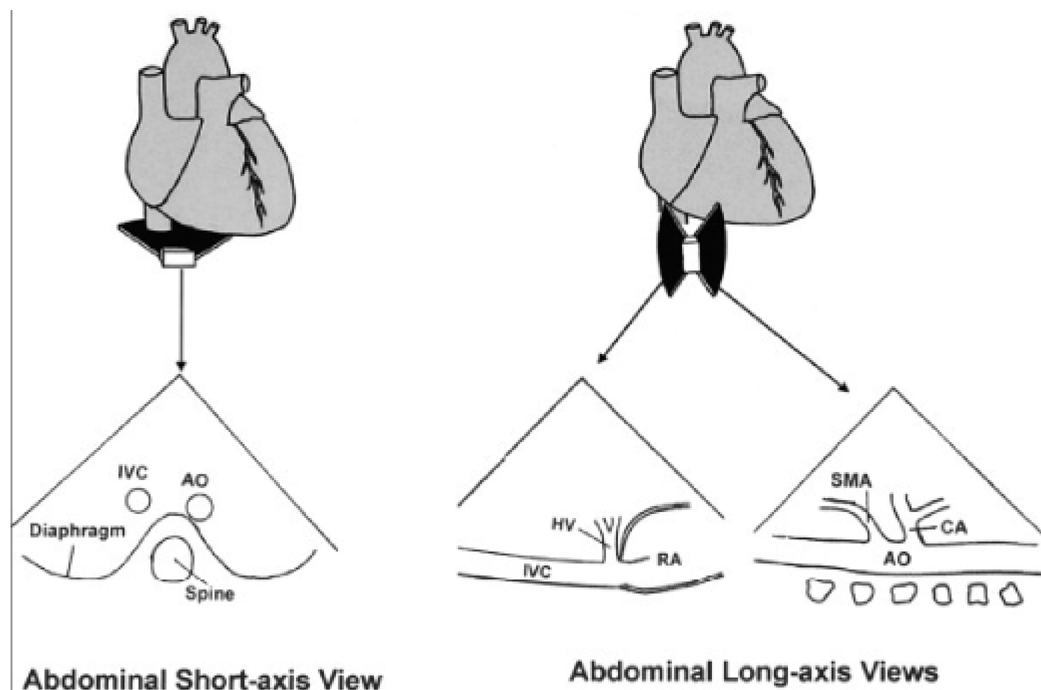
B, View showing the RV, RVOT, and pulmonary artery.
Subxiphoid Short Axis View RVOT/PV Plane

- *Transducer located just below xiphoid process.*
- *Indicator is at 6:00.*
- *Tilt ultrasound beam even more leftward.*

C, Short-axis view of the ventricles.
Subxiphoid Short Axis view LV Plane

- *Transducer located in just below xyphoid.*
- *Indicator is at 6:00*
- *Tilt ultrasound beam to patient far left shoulder.*

Azg. V, azygos vein; LA, left atrium; LV, left ventricle; MV, mitral valve; PA, pulmonary artery; RA, right atrium; RPA, right pulmonary vein; RV right ventricle, SVC,superior vena cava, TV, tricuspid valve.

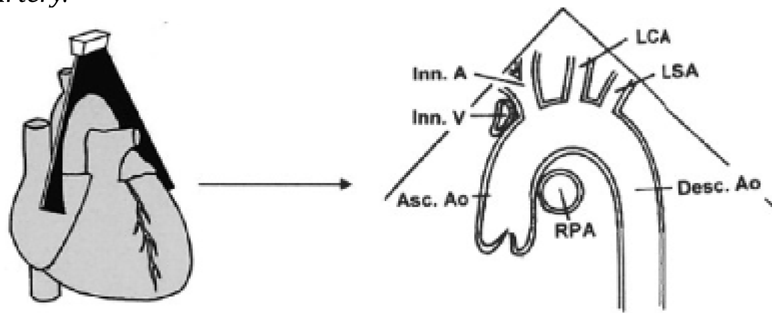


Abdominal views.
Left, abdominal short-axis view.
Abdominal View
Short axis view
Transducer located abdomen
Indictor is 3 00
'Ao on pts left.lVC on right

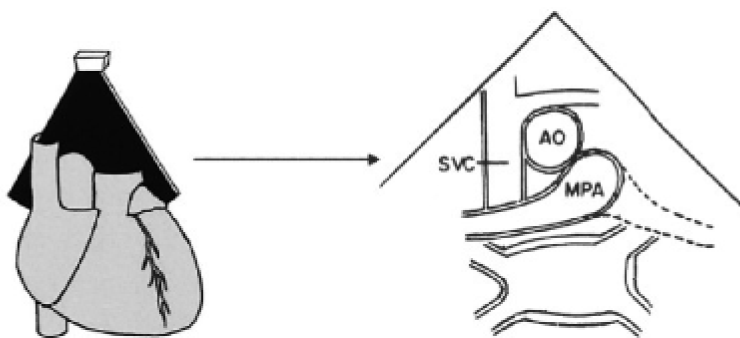
Right, abdominal long-axis views.

- ü *Abdominal View(AOSMA)*
  Transducer located in the abdomen 'Indicator is at 12 00
  leftward angulation of transducer
- ü *Abdominal View(IVC)*
  Trancducer located in the abdomen
  'Indicatoris at 12:00 ' Slight nghtward

AO, aorta; CA, celiac axis;HV, hepatic vein; IVC, inferior vena cava; RA, right atrium; SMA, superior mesenteric artery.

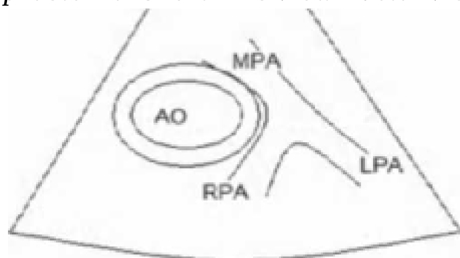


Suprasternal Long-axis View



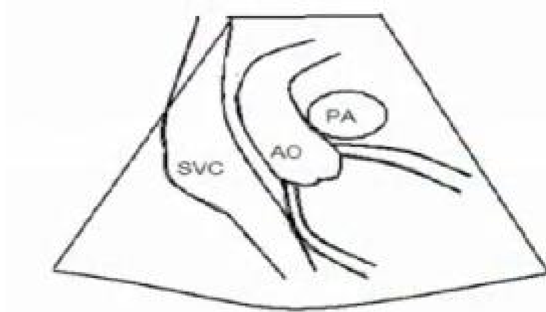
Suprasternal Short-axis View

Diagram of suprasternal notch two-dimensional echo views.
Top, Long-axis view.
Suprasternal Long-Axis View
Transducer placed in the suprasternal notch
Indicator towards 12:00
Tilt patient's chin left or right as needed
Bottom, Short-axis view(Crab View)
Suprasternal Short-Axis
•Transducer placed in the suprasternal notch
•Inducator towards 3:00
•Anterior tilt
Suprasternal Short-Axis View Ductal View



- *Transducer placed between Suprasternal notch and high left Sternal boarder*
- *Indicator towards 2:00-3;00*
- *Bifurcation of PA can be visualized.*

Suprasternal Short-Axis View Anterior Tilt



- *Transducer placed in the suprasternal notch*
- *Indicator towards 3:00*
- *Anterior tilt*

AO,aorta; Asc. Ao, ascending aorta; Desc. Ao, descending aorta; Inn. A, innominate artery; Inn. V, innominate vein; LA, left atrium; LCA, left carotid artery; LSA, left subclavian artery; MPA, main pulmonary artery; PA, pulmonary artery; RPA, right pulmonary artery; SVC, superior vena cav

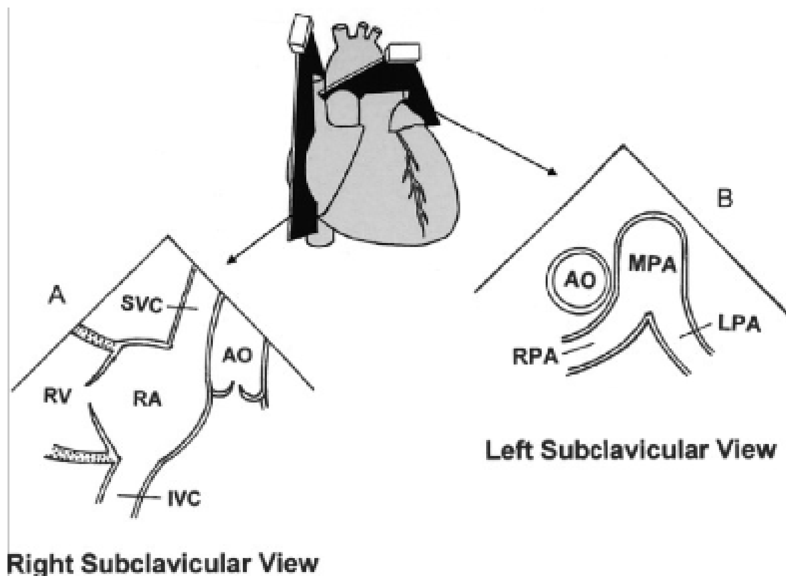
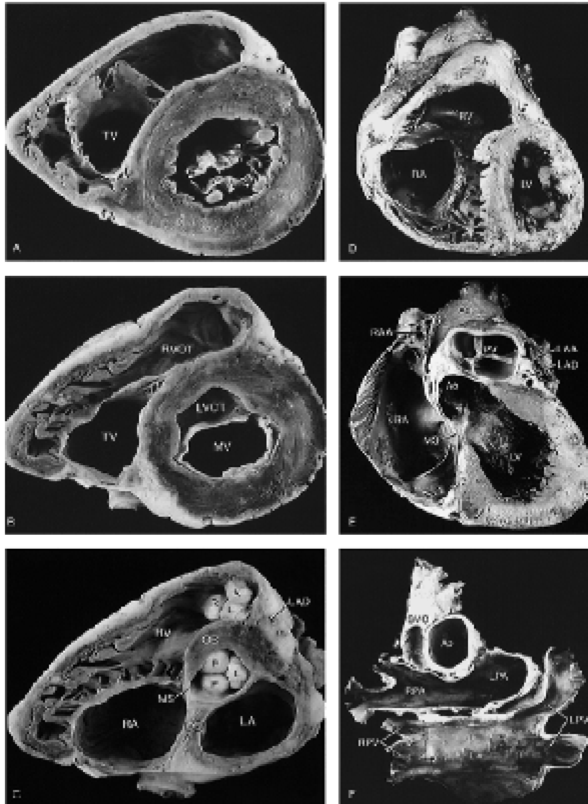
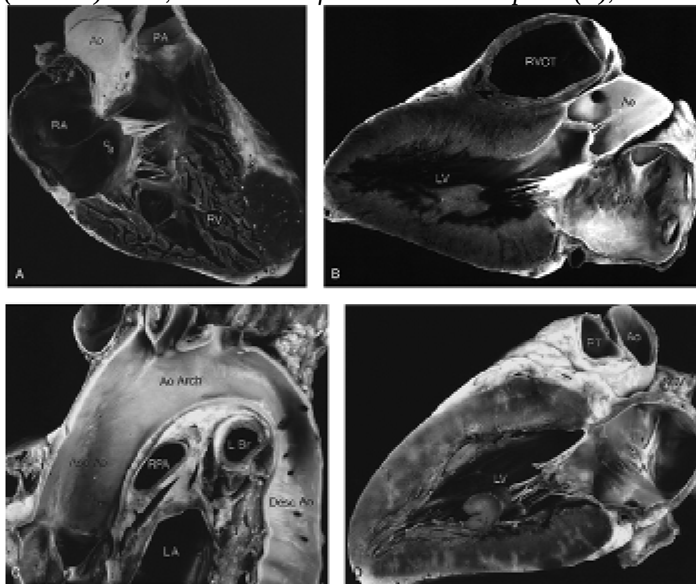


Diagram of subclavicular views. A, Right subclavicular view. B, left subclavicular view. AO, aorta; IVC, inferior vena cava; LPA, left pulmonary artery; MPA, main pulmonary artery; RA, right atrium; RPA, right pulmonary artery; RV,right ventricle; SVC, superior vena cava.

Topographic views:

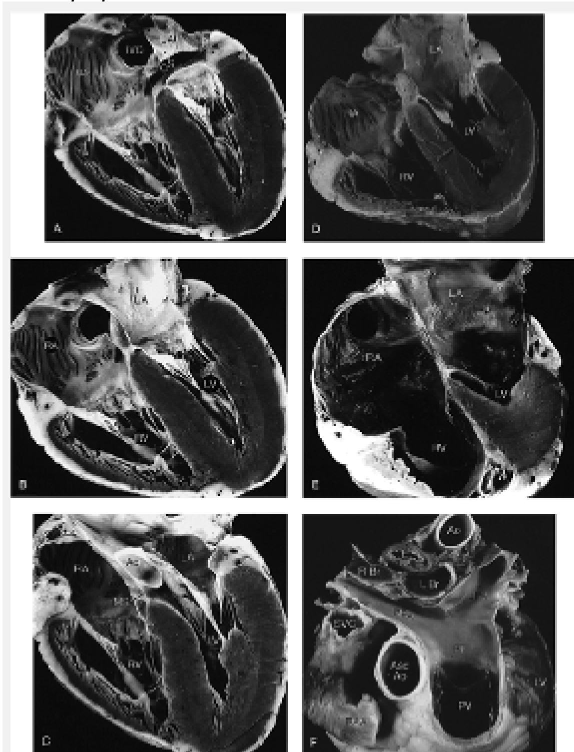


Tomographic methods of cardiac dissection (short-axis and frontal views), shown in normal hearts. A to C: Short-axis views, at the levels of the mitral valve orifice (A), left ventricular outflow tract (B), and aortic valve (C). D to F: Frontal (coronal) views, at the levels of the ventricular septum (D), membranous septum ((E), and left atrium (F).



Tomographic methods of cardiac dissection (long-axis and two-chamber views), shown in normal hearts. A and B: Long-axis views show inflow and outflow tracts of right ventricle (A) and left ventricle (B). C: Long-axis view of thoracic aorta

shows left bronchus and right pulmonary artery traveling beneath aortic arch. D: Two-chamber view demonstrates inflow tract of left ventricle.



Tomographic methods of cardiac dissection (four-chamber and horizontal views) shown in normal hearts. Aâ€“C: Four-chamber views, at levels of coronary sinus (A), fossa ovalis (B), and aortic valve (C). D to F: Horizontal (transverse) views at levels of ventricular inflow (D) and outflow (E) tracts and pulmonary artery (F).

Optimizing the Doppler Examination

§ The robustness of Doppler echocardiography as a tool for evaluating cardiac physiology is manifested only when its practitioners exhibit precise and diligent technique.

§ Doppler spectral envelopes need to be sharp and free of feathering. A sharp envelope is first achieved by aligning the ultrasound beam as parallel to the flow as possible.

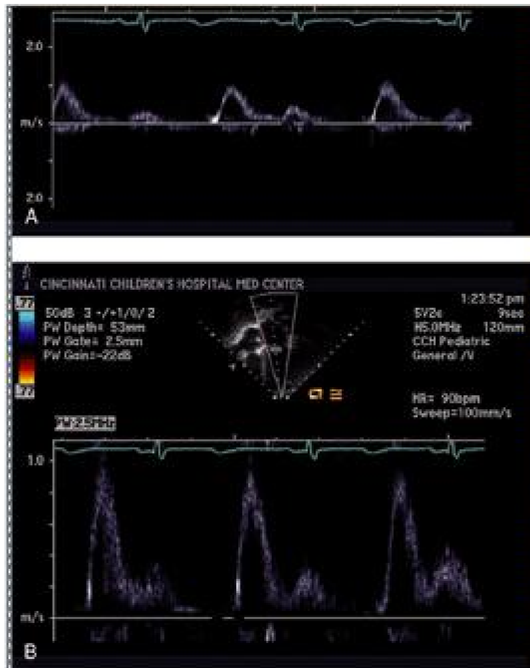
§ *Traditionally, color Doppler is used before application of pulsed or continuous wave Doppler* to determine the precise location and direction of a jet.

§ The transducer position on the chest is then moved accordingly so that the flow is directed either exactly toward or opposite to it.

§ The best transducer position for the Doppler examination may therefore be offset from the most ideal position for two-dimensional imaging.

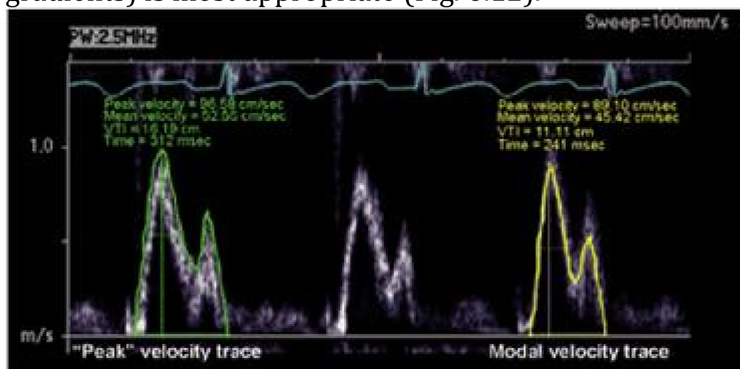
§ Second, the practitioner must be careful to *avoid overgaining* the spectral display, which can cause indistinct envelopes.

§ Third, *the spectral display of interest should fill as much of the screen* as possible by shifting the baseline up or down and decreasing the Doppler scale. In this way, the envelope is made as large as possible, minimizing the effect of imprecise Doppler envelope planimetry .



Mitral valve Doppler spectral profile recorded with (A) inappropriate and (B) appropriate scale and baseline shift. It is important to fill as much of the Doppler display area, thereby enlarging the Doppler spectral profile as much as possible, thus minimizing error in measurement (B).

§ Fourth, *tracing the Doppler envelope needs to be careful, precise, and steady*. The operator must know if a trace of modal velocity (for continuity equation) or of peak velocity (for pressure gradients) is most appropriate (Fig. 6.12).



Mitral valve Doppler spectral profile demonstrating the difference between a peak velocity trace (green) and modal velocity trace (yellow). The peak velocity trace follows the outside edge of the Doppler spectral contour whereas the modal velocity trace follows the midline of the contour. Peak velocity traces are used to obtain pressure gradients across valves and orifices. Modal velocity traces are used in the continuity equation and in calculation of cardiac output. Note that there can be a potentially large difference in the values obtained from a peak and modal velocity trace. (e.g., 31% difference in the two velocity time integrals [VTI] in this case).

§ Finally, the practitioner must be aware of the various Doppler modalities available to help optimize the spectral signal.

Color Flow Doppler

§ The color Doppler modality interrogates flow with multiple pulsed Doppler sample volumes placed successively along multiple scan lines. For each sampling gate, the baseline frequency is

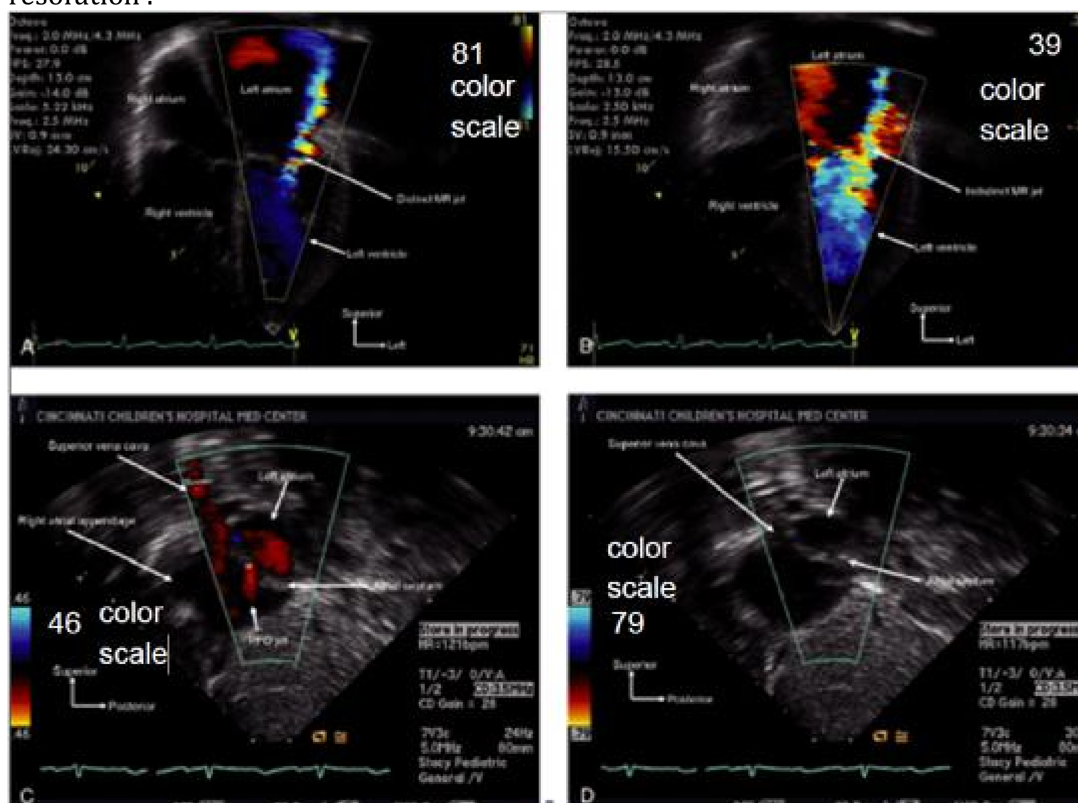
compared with the received frequency. Pixels in the image are assigned a color (red for flow toward the transducer and blue for flow away from the transducer) and a color intensity based on the magnitude of the *mean velocity*.

§ The color *Doppler scale* should be actively manipulated throughout the examinationion using *low-velocity* scales when interrogating venous flows (e.g., antegrade atrioventricular valve, atrial shunt, cavopulmonary, and systemic and pulmonary venous velocities) or *flows generated from low-pressure gradients* and *high-velocity scales* when interrogating arterial flows or flows generated from high-pressure gradients.

§ The examiner must actively think about and anticipate expected physiology during the study so that the color scale is appropriately adjusted .

§ For example, in a toddler being evaluated for a suspected ventricular septal defect, interrogation of the septum with a high color Doppler scale is appropriate. However, using such a high color scale in the investigation for a ventricular septal defect in a newborn would likely miss the shunt flow since the shunt is driven by a very low pressure gradient owing to the normally elevated pulmonary vascular resistance in the newborn period. The examiner would need to interrogate the ventricular septum with a low-velocity color Doppler scale in this instance.

§ These flows then need to be more carefully and precisely interrogated and quantitated with either pulsed or continuous wave Doppler. Because of the massive amount of data, a color *Doppler sector should be kept as narrow as acceptable* to improve accuracy and/or temporal resolution .



Series of four images demonstrating inappropriate and appropriate color aliasing limits during the echocardiographic examination.
Throughout an echocardiographic examination, the echocardiographer must actively decrease and increase the color aliasing limit depending on the expected velocity of the color jet being interrogated. For example, in evaluating high-velocity jets such as mitral regurgitation (MR), color aliasing limits should be high(high scale) as in the apical four-chamber view (A). This provides a clean, crisp and interpretable display of the mitral

regurgitation flow. Inappropriate low color aliasing limit (B) produces an indistinct display of the mitral insufficiency jet. On the other hand, when evaluating a low-velocity color jet such as a patent foramen ovale (PFO), the color aliasing velocity should be lower(low scale) to enhance recognition and display as in the subcostal image (C). If the aliasing velocity is set too high, detection of the low-velocity PFO jet may be impossible (D).

High flow → *high scale velocity* → *high color aliasing*
Low flow → *low scale velocity* → *low color aliasing*

Pulsed Wave Doppler

- Pulsed wave Doppler, along with continuous wave Doppler, is the principal echocardiographic tool for evaluating cardiovascular physiology.
- Pulsed Doppler causes the transducer to alternately transmit and receive short ultrasound bursts.
- The time between transmission and reception allows calculation of the depth of the signal or range-gating, which provides the operator with the Doppler frequency shift at a specific location.
- A disadvantage with the technique is that the maximal detectable frequency shift is limited by the Nyquist limt. However, the Nyquist limit can be extended by shifting the baseline of the spectral display, exchanging to a lower-frequency transducer, or moving to a different imaging plane so that the structure of interest is at a shallower depth if possible.
- High-pulse repetition frequency is a technique in which volleys of pulses of ultrasound are sent before reception of prior pulses. This technique increases the Nyquist limit but causes some range ambiguity.

Continuous Wave Doppler

- With the continuous wave Doppler modality, the transducer is continuously transmitting and receiving ultrasound signals.
- The disadvantage of this process is the absence of range-gating, but a major advantage is that the sampling rate is infinite so there is no longer a limit to the maximal frequency shift.
- The spectral display consists of a composite of signals with the maximal velocity representing the peak velocity at any depth in the plane of the ultrasound beam.

Appendex2

Normal values

M-MODE			
Normal Values	BSA (m2)	Mean (mm)	Upper limit (mm)
LVID	0–0.5	24	32
	0.5–1.0	34	40
	>1.0	40	48
RVID	<0.50	12	18
	0.5	10	18
	>1.0	12	18
LVPW	<0.5	5	6
	0.5–1.0	6	8
	>1.0	7	8
LA	<0.5	17	24
	0.5–1.0	21	28
	1.0–1.5	24	32
Aorta	<0.5	12	16
	0.5–1.0	28	22
	>1.0	22	28

Doppler Echocardiography

Doppler velocity values in Children

Velocities	Range	Mean
Tricuspid valve	0.3–.8 m/sec	0.6
Mitral valve	0.8–3.0 m/sec	1.0
Aortic Valve	1.0–8.0 m/sec	1.5
Pulmonic Valve	0.7–2.0 m/sec	1.0

Doppler gradient

PS/AS/Coarctation of aorta can be assessed by doppler gradient across valve or obstruction.

■ PS/AS Severity—Assesment

• Gradient >15 mmHg—abnormal

• Upto 50 mmHg—mild

• 50–7 5 mmHg—moderate

• >75 mm Hg—severe

■ Coarctation gradient >30 mm Hg significant

Mitral valve-Tricuspid valve

It is assessed by measuring:

■ Valve gradient (peak; mean)

■ Valve area indirectly (pressure half time)

• MS: Normal MVA-4–6 cm2/m2

• Mild MS-Above-1.5 cm2/m2

• Moderate MS-1–1.5 cm2/m2

• Severe MS-<1 cm2/m2

• TS-SevereTS-<1.3cm2

■ Right arm systolic BP–IV gradient=RVSP=PA pressure

■ Normal PA pressure = 25–30 mmHg

• Mild	PAH 30–50 mmHg
• Moderate	PAH 50–75 mmHg
• Severe	PAH > 75 mmHg

Tricuspid Regurgitation

■ Peak gradient +10 mmHg=RVSP (Fig. 3.5)

■ PR gives PA diastolic pressure

■ Trivial PR is physiological

■ PR is pressure difference between PA and RV in diastole

■ RVEDP+PR jet = PA diastolic pressure

■ Normal PA diastolic pressure is 10 to 15 mmHg

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Internet Based Echo Data

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